European Journal of Medical and Health Sciences
Vol 6 | Issue 6 | November 2024
ISSN 2593-8339

CASE STUDY

Tumors of the Skull Vault in Three Cases and a Systematic Review of the Literature

Liévin Panu, Salma Lahlou*, Abdelmajid Chellaoui, Khadija Ibahioin, Abdessamad Naja, and Abdelhakim Lakhdar

ABSTRACT

Tumors of the vault of the skull are rare and truly diverse, dominated in frequency by secondary lesions. Data from the literature concerning the frequency of bone tumors and the cranial location of primary tumors vary from 0.8% to 2%. They can be incidentally discovered or revealed by local signs, the most frequently encountered being painful or painless cranial deformities. The clinic is generally not very suggestive of the tumor specifically in question, although it can guide through the signs and symptoms present (such as inflammation and pain). The neuroradiological assessment, essential for their management, is no longer conceivable without computed tomography. We have collected in this work, in the light of a retrospective study including 3 cases, the pathogenetic, epidemiological, clinical, diagnostic, and therapeutic aspects within the Neurosurgery Department of the CHU Ibn Rochd of Casablanca of the tumoral lesions at the level of the vault of the skull. The etiological diagnosis of vault tumors is very polymorphic. We collected 3 cases whose average age was 30 years, with extremes ranging from 12 to 45 years with a female predominance, and whose histological diagnoses were all different from each other, others.

Keywords: Bone, skull, tumors, vault.

Submitted: July 21, 2024

Published: November 23, 2024

10.24018/ejmed.2024.6.6.2176

Department of Neurosurgery, Ibn Rochd University Hospital, Morocco.

**Corresponding Author:*
e-mail: drlahloualmal@gmail.com

1. Introduction

The skull vault, or calvaria, is formed through a process of membranous ossification and is composed of several bones: the frontal bone, parietal bone, temporal bone scale, lamina ascendens of the sphenoid, and interparietal bone. Its primary role is to protect the intracerebral organs from external harm, highlighting its importance [1].

When X-rays are taken from a cranial or cerebral perspective, swellings within the skull vault are often observed. Tumors of the cranial vault, although rare, have limited clinical manifestations but widely varying etiologies. These tumors can be primary or secondary, benign or malignant, and may either develop strictly within the bone or extend intracranially. This text presents the clinical and radiological characteristics, the specifics of surgical management and adjuvant treatment, and the prognosis for each tumor type.

While the embryological origin of the skull vault bones remains a subject of debate, a thorough understanding of embryology and normal anatomical variations is crucial for studying these tumors. Neuroradiology has advanced significantly in recent years thanks to increasingly efficient investigative methods, allowing for the development of management algorithms.

Given the wide range of etiologies, a comprehensive etiological review is necessary, with a focus on the specific characteristics of each. Therapeutic approaches may involve surgery, radiotherapy, chemotherapy, or a combination of these modalities. In rare cases, treatment may be abstained from, with the issue being purely aesthetic.

2. Materials and Methods

Our study aims to highlight the characteristics of different bone tumors of the skull vault through a series of three patients diagnosed and operated on at the Neurosurgery Department of the Ibn Rochd University Hospital in Casablanca over a 10-year period from March 2011 to February 2021. We conducted a bibliographical review focused on tumors specifically located in the cranial vault, supplemented by descriptions of the typical characteristics of tumors that may occur in this area.

The research was carried out at the Faculty of Medicine and Pharmacy of Hassan II University of Casablanca,

various university medical libraries, and the Ibn Rochd University Hospital. The three patients included one male and two females, aged between 12 and 45 years. Symptoms were primarily characterized by headaches and the presence of a hard, painful subcutaneous mass. Diagnostic exploration was mainly conducted using CT and/or brain MRI. A significant challenge we faced was the rarity of tumors located in the skull vault.

3. Discussion

The cranial vault refers to the membranous bones, including the frontal, parietal, temporal, and occipital scales, in the region above a line that runs through specific anatomical landmarks such as the nasal notch, the orbital rim, the external orbital process of the frontal bone, the infratemporal crest of the sphenoid, the upper portion of the external auditory canal, and the posterior edge of the occipital foramen [2]. Some authors consider the posterior limit to be the upper occipital curved line. These bones are connected to each other by synarthrosis or sutures. In newborns, membranous spaces not yet calcified are individualized between the bones of the skull, known as fontanelles, of which there are four: Bregma, Lambda, Pterion, and Asterion. These spaces become totally ossified around the age of 2. This work does not delve into the descriptive anatomy of the skull vault and its embryological aspects.

Generally, tumors located on the skull vault are rare. Data from the literature concerning the frequency of bone tumors and the cranial location of primary tumors vary from 0.8% to 2%. These tumors are mainly dominated by secondary lesions. For example, in the Vanderberg series, primary cranial tumors account for only 1.4% of all primary bone tumors. The study by Meder *et al*. [3] includes secondary tumors, such as metastases (e.g., thyroid metastases), bone localizations of hemopathies (e.g., KAHLER disease), and skull damage by neighboring tumors, particularly meningiomas, which is consistent with our study.

Cranial vault tumors affect both children and adults. In children and adolescents, the most common are dermoid and squamous cell cysts, and they are also secondary to neuroblastoma metastases [4]. In adolescents, osteoma is more common. However, in adults, various origins are seen, with breast and lung cancer metastases being the most dominant [3]. In our series, the average age of patients was 30 years, with ages ranging from 12 to 45 years.

No general predominance of one sex has been noted; the preponderance depends on the tumor itself. For instance, meningiomas and hemangiomas are more frequent in women, while Paget's disease and Kahler's disease predominate in men, as proven in our study. Several studies report parietal localization, which is consistent with our series.

During our study, we did not find a preferred site for tumors of the cranial vault (see Table I). Metastases, the most frequent of these tumors, are generally multiple. Primary tumors such as squamous cell cysts tend to have a preferential frontal, parietal, or occipital location. Apart from the embryological origin of certain tumors, others are favored by trauma and radiotherapy, and sometimes the genetic origin is found.

Although the sample size in our series is very small due to the rarity of the pathology treated, we aimed to create an illustrative table by comparing our findings with the data in the literature. Tumor lesions of the cranial vault can be discovered incidentally during an assessment for unrelated conditions [4], or they may be revealed by local signs, primarily cranial deformities, which may or may not be painful. In rare cases, symptoms may include nerve compression, epileptic seizures, dizziness, and signs of intracranial hypertension, all of which typically progress gradually [4].

Clinical examination provides limited information unless swelling is present, in which case the characteristics such as hardness, renitence, fixity, mobility, and the condition of the subcutaneous plane are evaluated [4]. Consistency may suggest diagnostic hypotheses [5], but it is almost impossible to clinically determine the relationship between the tumor and the bone [5]. Differential diagnosis should consider congenital cranial dysmorphisms such as trigonocephaly, plagiocephaly, or post-traumatic dysmorphia.

The skull vault comprises an internal and external lamina, with the diploe—a spongy bone layer containing diploic veins—situated between them. These veins, which have thin walls and no valves, contribute to a rich craniocerebral anastomotic system that provides collateral circulation during venous sinus occlusion and serves as a pathway for the spread of infections through the vault.

Computed tomography (CT) and magnetic resonance imaging (MRI) are the primary diagnostic tools. Traditional skull X-rays play a minimal role today, having been largely replaced by CT scans. Non-invasive or minimally invasive angiography, using either CT or MRI, is reserved for specific diagnostic indications or therapeutic intervention procedures. While MRI provides superior anatomical detail, both MRI and CT are essential in clinical practice. CT allows for the study of the relationship between bone abnormalities, soft tissues, and the underlying brain, providing details about tumor characteristics such as boundaries, matrix appearance, calcifications, and involvement of internal and external tables [3]. MRI, though limited in bone studies, offers superior three-dimensional analysis, particularly in understanding the tumor's relationship with brain structures and dura mater sinuses [3].

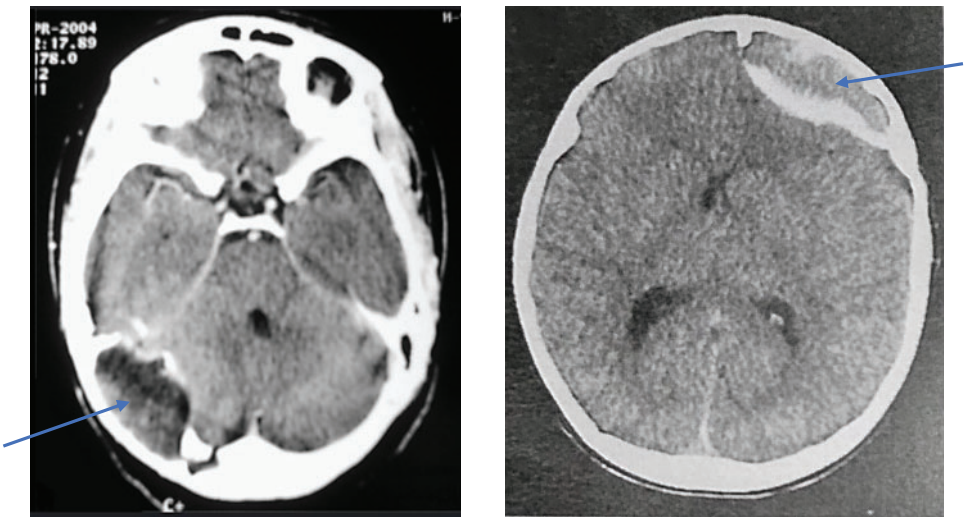
Scintigraphy, although nonspecific, remains the reference examination for detecting metastases due to its high sensitivity, retaining its value in identifying benign or pretumoral lesions [3]. The cranial vault is vascularized by branches of the middle meningeal artery and is traversed by venous sinuses. Angiography, guided by the tumor's location, is mainly used for intraoperative embolization of highly vascularized tumors to reduce bleeding during excision [3], though its diagnostic role is now limited [3]. Ultrasound is not used in adults because the acoustic windows are no longer available as the fontanels are closed. However, it is valuable in infants, where its lack of irradiation allows for close monitoring.

The discovery of a bone abnormality on standard skull images, performed with or without a cranial call point, should always prompt a CT scan. This non-invasive examination, using appropriate windows, facilitates a relatively easy study of the bone, soft tissues, and adjacent brain.

TABLE I: Epidemiology of Cranial Vault Tumors in Our Series according to Sex, Age, Tumor Site, Frequency, and Postoperative Evolution

Tumors	Sex	Preferred age	Preferred tumor site	Frequency	Postoperative evolution
Grade 1 benign meningioma	M < F	50–60 years old	Pterional	Not specified in the literature	Good evolution without recurrence
Metastasis of papillary thyroid carcinoma	M = F (=according to the literature)	Not specified	Multiple	More common tumors of the cranial vault [3]	Not specified in the literature Recurrence (our series)
Epidermoid cyst	M > F (=in literature) [1], [2]	20–60 years old	Pterional and coronal suture proximity	Not specified in the literature	Good evolution without recurrence

Note: F: Female, M: Male, >: Superior, <: Inferior, =: Equal.



(a) (b)

Fig. 1. Brain CT axial sections showing (a) a heterogeneous cystic mass with hypo and left frontal hematoma extra dural in the hyperdense areas and (b) the lesion-destroying process of calcification (blue arrow). The left occipital bone (blue arrow).

The etiologies of skull vault tumors vary depending on the patient's age. We have primary tumors, including:

- Primary tumors of embryonic origin (dermoid cysts, epidermoid cysts, and chordomas),
- Tumors of bone origin (osteomas),
- Primary tumors of vascular origin (hemangiomas and aneurysmal cysts),
- Tumors of fibrous origin (fibroid ossificans, fibrosarcoma, and fibrous dysplasia),
- Giant cell tumors,
- Ewing's sarcoma,
- Primary reticulosarcoma (from PARKER and JACKSON),
- Cartilaginous tumors (chondromas and chondrosarcomas).

The epidermoid and dermoid cysts, formerly called pearly tumors or cholesteatomas [6], have an estimated frequency between 0.3% and 2% of primary intracranial tumors [7]. Their pathogenesis is a subject of several controversies, mainly between the congenital theory, which involves ectodermal embryonic debris located in the diploe [8], and the acquired post-traumatic theory, such as a penetrating wound [1], [8]. Nevertheless, dermoid cysts are always of embryological origin, with their preferred location being endocranial [8].

Epidermoid cysts are benign lesions with slow progression, usually discovered in adults between the ages of 20 and 60, with an average age of 34 years [7] and no predominance of sex [1], [5]. Prenatal discoveries may also exist. Originally, in the diploe, the cyst pushes back the external and internal table, giving rise to a soft swelling, sometimes renitent and not painful. A neurological syndrome of compression can occur when the extension is intracranial [8]. The invasion can be both endocranial and exocranial, resulting in an hourglass tumor [8]. In the case of multiple recurrences, malignant transformation is possible but rare.

The preferred site of epidermoid cysts is the pterional region and the lateral parts of the coronal suture, though various other locations are possible [9]. The preferred sites are the frontal and parietal bones, according to Skandalakis [5]. The radiological diagnosis of epidermoid cysts is suggested in the presence of a rounded lacunar lesion with clear boundaries, surrounded by a continuous peripheral sclerotic reaction. The lacuna has homogeneous transparency, but sometimes the base is areolar, with the presence of partitions and a cloudy or foamy cauliflower appearance [8].

A CT scan reveals the diploic site of the lesion, blowing out the two tables due to its extension and sometimes destroying them, making intracranial invasion possible (see Fig. 1). The squamous cell cyst has a density lower than or identical to the parenchyma, not modified by the contrast medium. On MRI, the signal is variable, depending on the chemical composition. The angiography appearance is nonspecific, showing an extracerebral

avascular process [1]. Histology remains the only means of confirming the diagnosis.

Epidermoid cysts are differentiated from dermoid cysts of ectodermal and mesodermal origin by their rather median and paramedian position on the vault, the presence of cutaneous appendages (sebaceous glands, sweat glands, and hair follicles) in the cystic membrane, a hypodense content that is frankly negative on CT scan, and parietal or central tumor calcifications with a hyperintense appearance in T1 and a relatively hypointense appearance in T2. These lesions are sometimes difficult to differentiate from a cholesteatoma developed because of a middle ear infection. The radiological appearance of dermoid cysts is substantially identical to that of squamous cell cysts, with the density being heterogeneous due to the coexistence of elements of different origins.

Surgery is the treatment of choice, as the excision must be complete to avoid any recurrence. They generally have a good prognosis after complete excision [1].

Osteomas are benign, slow-growing tumors most often discovered during the 4th and 5th decades [3] but can be observed at any age. Their preferred age is between 20 and 50 years, according to some authors [10]. Their prevalence is unknown due to their asymptomatic nature. The pathogenesis is controversial [10]; three theories are described: embryological, infectious, and traumatic [10]. Osteomas of the cranial vault are less common than those of the skull base. The exostic form, originating from the external table (exostosis), is more common than the enostic form, which originates from the internal table (enostosis) [11], [12].

Generally solitary, they are mainly located in the paranasal sinuses, particularly the frontal sinus [3]. On the cranial vault, they most often develop from the external table (exostosis) [11]. They are generally of parietal location [3] and are manifested by localized swelling of the same consistency as the neighboring bone [3], lasting without pain [4]. Osteomas developed from the internal table (enostosis) are exceptionally symptomatic. Multiple locations, which are rare, may be part of Gardner's syndrome [3]. The differential diagnosis is made with an osteochondroma or an ossifying fibroid [13]. Asymptomatic osteomas do not require treatment except in the case of rapid and extensive growth. Simple excision is the treatment for those who are symptomatic [14].

Osteosarcomas are tumors that arise from osteoforming connective cells. These are malignant tumors usually affecting older children or young adults [15]. 75% of patients are between 10 and 25 years old. They are very rarely found before the age of 5 and after the age of 30 years [16]. The location on the cranial vault represents 0.7% to 3% of all locations, according to some authors [17]. They are the most common primary bone tumor after myeloma [15]. They can also be secondary, as is the case following Paget's disease or radiotherapy [18], especially after radiation in pediatric age.

The clinical sign is mainly pain. The general condition is maintained for a long time and subsequently deteriorates with asthenia and hyperthermia [8]. Differential diagnosis is often difficult in radiology and sometimes impossible in histology. Osteosarcoma rarely causes lung metastases [19]. The seat is primarily the posterior convexity of the vault [20]. Biologically, anemia and leukocytosis can be observed, and sometimes there is an increase in alkaline phosphatase [8].

CT scans with contrast agent injection reveal hypervascularization, tumor calcifications, bone destruction, and especially exocranial and/or endocranial invasion [8]. Angiography shows a hypervascularized **tumor** [4] with dilated pedicles and the existence of arteriovenous shunts, also visible on MRI [8], [20], [21]. Surgery is desirable if the dural sinuses are not invaded [4]. Its prognosis has been improved with the introduction of multidrug therapy in various protocols [19]. Maurizio Salvati *et al*. [16] are convinced that chemotherapy should be started as soon as evidence of osteosarcoma has been established, i.e., before surgical resection and immediately after biopsy examination. They believe that radiotherapy should be restricted to inoperable cases or with postoperative tumor residues. Osteosarcoma juxtacortical differs from the previous form by its slow evolution, and its prognosis is more favorable [3]. It is poorly vascularized [4] except in the case of dedifferentiation [3], making surgery difficult.

Hemangiomas are tumors of vascular origin; they constitute about 1% of symptomatic primary bone tumors. This figure is certainly underestimated, as evidenced by the frequency of fortuitous discoveries [3]. These are benign lesions, though their tumor nature is disputed. They are exceptionally found in the context of angiomatosis [22]. They are found mainly in adults between the 4th and 5th decades of life, with a clear female predominance [3], particularly during genital activity [8]. The cranial vault, vertebral bodies, and ribs are the main locations of these lesions [3]. They develop everywhere on the vault, with the frontal and parietal regions being the most frequently affected [8], [19]. Hemangiomas usually manifest as a soft, painful swelling [4]. Their sensitivity is particularly noticeable during menstruation or pregnancy [8]. Their consistency is often hard and becomes renitent or soft when the outer table has been destroyed [8].

A CT scan confirms the diploic site of the lesion, with the lifting of two tables usually respected. It also shows the characteristic appearance of a "honeycomb" or "sunbeam" [4]. On MRI, contrast is usual, and foci of fatty hypersignal may be found [23]. In this category of tumors of vascular origin, there are also aneurysmal cysts, which we will not discuss here. As the list of tumors of the cranial vault is large, we will not cover them all in this work.

Ewing sarcoma is a malignant tumor of spinal cord origin whose cranial involvement is rare, representing about 1% of locations [18], [24]. Its origin is most likely neuroectodermal [25]. Pritchard *et al*. [26], out of 229 observations, found only 3 cranial locations. The clinical and biological picture can suggest its diagnosis: young subjects between 10 and 25 years of age in 75% of cases, with the highest incidence between 5 and 13 years of age and a predominance in men (ratio 1.6:1) [8]; painful swelling, fever, anemia, hyperleukocytosis, and acceleration of sedimentation rate. The frontal and parietal convexities are preferential.

Usually, the radiological aspect is that of a lytic expansive process whose density is increased after injection of contrast medium. The external and internal tables are first blown out in an "onion skin" pattern and then disappear.

The evolution continues with the invasion of the adjacent tissues, with a hyperostotic reaction producing small, rounded spicules clearly visible on tangential images [8]. Here, although MRI allows a better study of the tissue part of the tumor, it has a limited role [27].

A radioisotope CT scan is essential to rule out lesions at another bone site. It is the examination of the operative specimen that confirms the diagnosis, and its treatment is surgical, coupled with radiotherapy and chemotherapy. Chondrosarcomas are generally slow-growing tumors whose malignancy can be difficult to confirm on criteria other than evolution and histology [28]. Their radiological appearance may resemble that of chondromas. Mesenchymal chondrosarcomas are rarer than the classic forms, are extraosseous in almost half of the cases, and develop mainly from the meninges [29]. Chondrosarcomas are responsible for irregular bone erosion or even punched-out lysis. Their preferred site is the fronto-parietal vault, and they are typically discovered in the 3rd decade of life. The differential diagnosis of chondrosarcomas can be made with primary osteogenic sarcoma or osteochondroma of the skull [30]. A few exceptional cases of chondroblastoma vault have been reported [31]. Their radiological appearance can be imposing for an aneurysmal cyst: a diploic, heterogeneous lacuna with clear boundaries, blowing the two tables.

Secondary tumors or metastases, although they can cause severe disability due to compression of the dural sinuses or nerve structures, are often asymptomatic in the skull. Hematogenous propagation is the most common cause of cranial arch tumors in adults [3], [4], [8]. All cancers can metastasize to the skull vault; however, primary tumors of epithelial origin are the most frequent [32]. In adults, breast and lung cancers are most often involved [3], followed by thyroid, kidney, and prostate cancers, as noted by Artico *et al*. [32] in a retrospective study of ten cases. The orbit and skull are the preferred secondary locations of neuroblastomas in children [3].

Biology reveals a significant increase in sedimentation rate and alkaline phosphatases, although less pronounced than in Paget's disease, hypercalcemia, and hypercalciuria in osteolytic forms. Finally, an increase in acid phosphatases is observed in prostate cancer. It is important to note that the same tumor can, in the same patient, give osteolytic or osteosclerotic metastases, usually to different bones [32]. The involvement is unique in only 10% of cases. In any case, the definitive diagnosis is histological from biopsy or tumor excision. The usual appearance on radiology is that of multiple rounded or oval gaps, without peripheral condensing reaction, with more irregular contours than in myeloma lesions. In MRI, the injection of contrast medium is not necessary for diagnosing large metastases, but it improves the detection of small intradiploic lesions [33]. However, an MRI provides better information than a CT scan on the degree of dural infiltration or invasion of a venous sinus [34]. A periosteal reaction is rarely found.

The treatment of tumors of the cranial vault is most often surgical, combined with new and/or adjuvant chemotherapy, sometimes, combined with radiotherapy, depending on the histological type of the tumor in question. We cannot address this aspect without referring to preoperative preparation (anesthetic preparation). Perioperative hemorrhage is statistically significantly associated with an increase in postoperative morbidity. The preoperative assessment of bleeding risk is an essential step during the anesthesia consultation to develop corrective strategies to reduce this risk. The pathophysiology of perinterventional hemorrhage is complex and is based on two components: the bleeding risk related to the surgical procedure and the risk related to the patient, in other words, the existence of risk factors specific to the patient exposing them to an additional risk of bleeding. These individual bleeding risk factors may be constitutional or acquired and may affect primary hemostasis or coagulation.

Treatment is often surgical, with indications of excision of the bony cranial vault and meningeal excision. When the tumor is perfectly mobile on the deep bone plane, the excision is performed above the periosteal plane while respecting the established safety margins. In case of reduced mobility or intraoperative observation of infiltration of the periosteum of the vault or erosion of the external cortex, bone milling to the diploe is proposed. In the event of confirmed bone invasion (CT), a resection of the cranial vault is proposed.

Operative steps are planned sequentially, focusing on the cutaneous, bone, dural, and cerebral levels. Depending on the case, the tissues can be preserved in many instances at the skin level. In cases of larger resection, local cover flaps (rotation flaps) or free pedicles (radial, latissimus dorsi) should be considered. In the event of an expected skin defect, the placement of subcutaneous expanders several weeks before surgery can allow for the gaining of enough skin surface to fill the defect.

On the bone plane, a defect whose size must be anticipated will occur. The entire pathological bone must be respected. The passing flap of the lesion is cut for convexity lesions. Reconstruction is done through various options, including autologous cranioplasty by doubling a contralateral cranial flap, iliac graft, or costal sampling. In the case of heterologous cranioplasty, several techniques or materials can ensure reconstruction (cement, metal grill, PEEK, silicone, hydroxyapatite; see Figs. 2 and 3). Some materials can be made on-site, while others require custom fabrication in advance and must be anticipated.

At the dural and cerebral levels, a resection of the dura mater may be associated. The defect must be reconstructed with autologous tissue (temporal fascia, fascia lata, epicranium, etc.,) or heterologous (synthetic) materials.

In the case of epidermoid cysts, the procedure involves removing the cyst to prevent recurrence [5], [35]. The tumor capsule, which constitutes the germinal part of the cyst, must also be addressed [1]. While some authors believe that any discovery of an epidermoid cyst warrants surgical excision to prevent complications, others suggest that surgery should be considered only in cases of aesthetic damage, significant tumor size, or neurological symptoms [3].

Immediate outcomes depend on three factors: the tumor's location (more superficial tumors have better prognoses), the multiplicity of tumors (which contraindicates

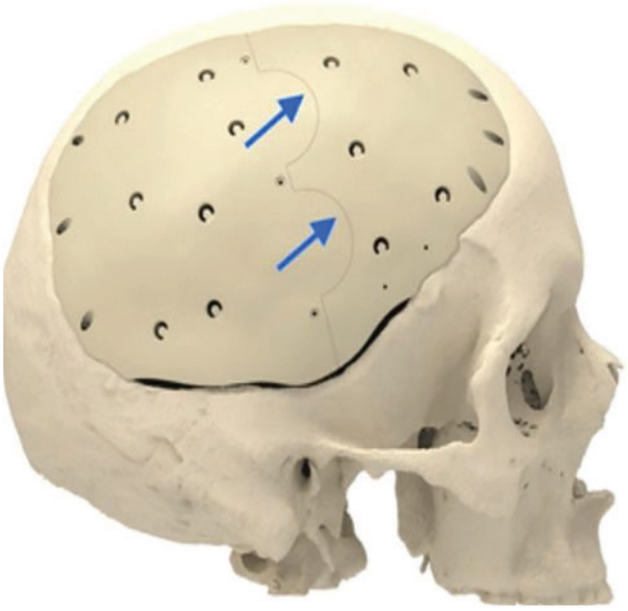


Fig. 2. PEEK implant.

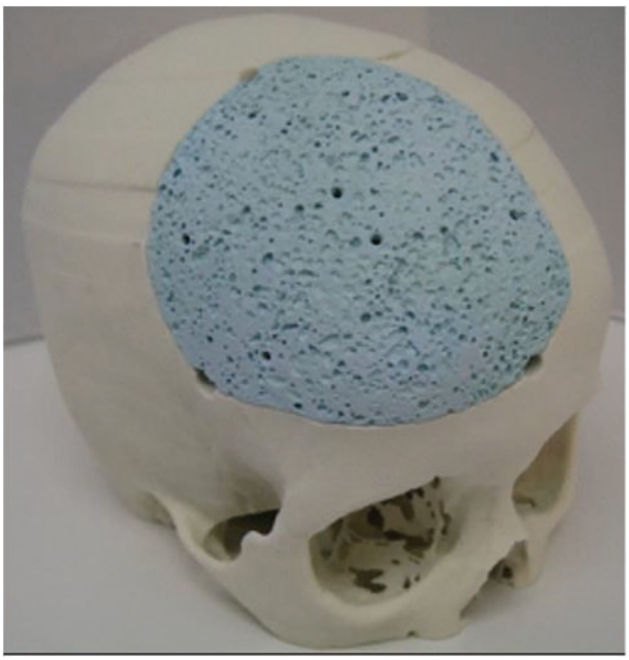


Fig. 3. Hydroxyapatite implant.

surgical therapy), and the tumor's histological characteristics (which affect the prognosis). The risk of recurrence is higher with incomplete resection of the tumor [9], [35], [36]. Depending on the patient's clinical condition, the extent of tumor invasion, and the histological findings, surgery may or may not be the sole therapeutic approach.

Complications, though not specific, include postoperative infection, cerebrospinal fluid circulation disorders, and decompensation of defects. Anti-convulsant therapy may be necessary for patients who experienced preoperative epileptic seizures. Specific complications can include cranioplasty mobility, postoperative hemorrhage, intracranial hypertension, and cerebral edema.

Systemic chemotherapy and targeted therapies are used for metastatic solid cancers or hematological conditions, while radiotherapy is applied to focal lesions, sometimes as an adjuvant to surgical resection. In some cases, embolization is required before surgery to reduce intraoperative bleeding, particularly when the tumor invades the superior longitudinal sinus, as is common with giant cell tumors [25] or certain meningiomas [35]. Radiotherapy is indicated alone or as a neoadjuvant or adjuvant treatment, but it can also be implicated in the malignant transformation of tumors like giant cell tumors [25]. It is useful in recurrences or inoperable meningiomas [32], [35], with solitary plasmacytoma being a radiosensitive tumor. Some authors recommend complete surgery without additional radiotherapy unless there is local recurrence.

Chemotherapy is particularly relevant for tumors like extracranial Ewing sarcomas, where survival rates have increased significantly with the introduction of adjuvant chemotherapy, from 8%–15% at 5 years to 55%–60% [26], [27]. Combination chemotherapy has revolutionized the prognosis of osteosarcomas since its inclusion in various protocols. Neoadjuvant and adjuvant chemotherapy is known to eradicate micrometastases, necrotize the tumor and its vascular ramifications, and facilitate

more conservative surgery [15], [16]. The therapies reduce the risk of local relapses and the number of metastases while extending the time to their development.

Therapeutic abstention is recommended for certain tumors, such as multiple tumors (e.g., metastases, hemopathies) and asymptomatic osteomas, except in cases of rapid and extensive growth [14]. Symptomatic osteomas require simple surgical excision [14].

We will briefly discuss multiple myeloma, solitary plasmacytoma, and lymphomas for cranial vault localization of hematological disorders. Pseudo-tumoral lesions (e.g., Paget's disease, histiocytosis X, sarcoidosis, hydatidosis) and cranial disorders of metabolic origin (e.g., hyperparathyroidism, dysvitaminosis) will not be covered in this work.

Multiple myeloma, or Kahler's disease, is a malignant plasma cell proliferation affecting bone and marrow, associated with a monoclonal immunoglobulin in the serum and urine. It is the most common primary bone tumor [8] and mainly affects individuals in their 5th and 6th decades. Cranial involvement occurs in about 50% of cases [3], [4]. Clinical examination is often unremarkable, initially focusing on joint pain and, later, on general health deterioration. Hemorrhagic syndrome, neurological syndrome (e.g., spinal cord compression), recurrent infections, and spontaneous fractures may also occur [8].

Myeloma of the cranial vault typically presents as multiple, disseminated, punch-shaped gaps without peripheral condensation [3]. Initially diploic, these lesions thin and eventually destroy the cranial tables [23]. Other observed aspects included seminated bone demineralization simulating osteoporosis, condensing lesions, and large locations sometimes causing cerebral compression [37]. Radiological features can resemble osteolytic metastases, but certain signs are more common in myeloma, such as clearer lacunar contours, symmetrical distribution, uniform size, and a homogeneous lacunar area [38]. Diagnosisisconfirmedthroughbiology and bone marrow biopsy, though radiological characteristics are recognized [4]. Sedimentation rates are elevated, and blood tests reveal blood cell

rouleaux, normochromic normocytic anemia, and hyperproteinemia with a beta or gamma immunoglobulin peak on electrophoresis identified [8]. Hypercalcemia and renal insufficiencies may also be present.

Solitary plasmacytoma is an isolated plasma cell malignancy without the clinical, biological, or radiological signs of myeloma [39]. With a better prognosis, it is less common than multiple myeloma and affects younger individuals under 50 years of age in half the cases. It affects both men and women, representing 2.5% of primary tumors and less than 4% of malignant bone tumors (). Its craniocerebral ocalizationis rare [7]. Preoperativediagnosisischallenging due to the nonspecific nature of the lesion.

MRI is useful for identifying the bone locations and assessing possible meningeal infiltration. Differential diagnosis includes multiple myeloma, aggressive meningiomas, metastases, bone hydatidosis, and chondrosarcomas [36]. There is no consensus on treating solitary plasmacytoma of the cranial vault [39]. While some treat it exclusively with radiotherapy, others argue that complete surgical excision makes radiotherapy unnecessary [39]. Chemotherapy has not shown effectiveness against the tumor [40].

Primarybonelymphomas are rare, accounting for 1%–4% of non-Hodgkin lymphomas. They are mostly found in adults but can occur at any stage in cases of immunosuppression. The Burkitt type is most common in children. Diagnosis is histological [41]. Bone involvement typically occurs in the disease and primarily affects the axial skeleton [41]. Malignant lymphomas of the cranial vault generally infiltrate and often appear as meningiomas on CT scans. Arteriography reveals visualization by meningeal vessels and provides information on the patency of the superior longitudinal sinus [42].

Neighboring tumors can reach the skull by contiguity or perineural extension. Meningiomas are benign tumors, usually discovered in women during the 5th and 6th decades. In imaging, meningiomas typically appear as expansive extra-axial processes with marked uniform contrast intake. They may be heterogeneous if cysts or necrosis are present, with calcifications in a quarter of cases and cerebral edema in 60% of cases [41]. Tumors can induce osteogenic reactions, with MRI superior to CT in analyzing vascular complications, especially venous sinus invasion. Histological analysis confirms the diagnosis, usually indicating a benign meningoepithelial or transitional tumor, though, in 5%–10% of cases, meningiomas are malignant [43].

In about 10% of cases, meningiomas involve the cranial vault and develop at suture height [41]. Intraosseousmeningiomais rare, originating from arachnoid cells in an ectopic position [3]. Malignant brain tumors can exceptionally destroy the cranial vault, as described during gene evolution.

Subdural and extradural intracranial metastases may extend to the vault, particularly from breast, lung, and prostate cancer [44]. Malignant soft tissue tumors can cause progressive osteolysis without inner table damage more pronounced than inner table damage. This occurs in squamous cell carcinoma, rhabdomyosarcoma, or malignant melanoma metastases. Obstetric accidents can generate extra-or intracranial collections in newborns, which may calcify over time.

Serosanguineoushumps, common in obstetric trauma, are localized blood suffusions in soft tissues, with the bone remaining intact. In adulthood, these traumas generally have no lasting effects. Calcifiedhematomas can form a calcareous shell, eroding the outer table. CT scans show a dissociation between this calcareous shell and the eroded outer table. Cephalhematoma, another obstetric trauma, results in a subperiosteal hematoma, typically varietal, limited by sutures and fontanelles. The underlying fracture line is visible, with calcification beginning on the 20th day. The diagnosis is based on history and characteristic radiological appearance. A few global cases of extradural hematoma calcification have been described, with calcium deposits and radiological images resembling subdural hematoma calcification. The diagnosis is based on history, absence of hypervascularization, and CT scan findings.

4. Conclusion

Tumors of the skull vault are rare, and several etiologies are recognized, with metastases at the forefront. Their clinical manifestation is, in most cases, a swelling of the cranial vault that may or may not be painful. Radiological examination, dominated by computed tomography, is essential for analyzing these tumors, including their exact location, extension, and relationships with the underlying tissues. Histology remains key to diagnosis, even though some tumors have quite specific clinical and radiological characteristics.

In the literature, there is no clear predominance for a given sex or a particular seat on the vault. However, the histology of tumors located on the skull vault shows that some are more often encountered in women than in men, such as meningiomas and hemangiomas. These tumors affect children, adolescents, and adults alike. There is a higher frequency of squamous and dermoid cysts in children and adolescents, in addition to osteomas in the latter group. In adults, the origin of these tumors is quite varied, with metastases from breast and lung cancers being the most common.

Their treatment is most often surgical, combined with neo-or adjuvant chemotherapy and sometimes radiotherapy, depending on the histological type of the tumor. Similarly, the prognosis is strongly linked to the specific type of tumor. Apart from the embryological origin of some tumors, others are caused by head trauma and radiation therapy. A genetic origin is sometimes found.

The discovery of tumor lesions in the cranial vault may be fortuitous during an assessment carried out in another context [4], or they may be revealed by local signs dominated by cranial deformities, which may or may not be painful. In rare cases, symptoms are due to the compression of nerve structures, leading to epileptic seizures, dizziness, and signs of intracranial hypertension. The symptomatology is generally progressive [4].

The clinical examination is not very informative, except in the case of swelling, where characteristics such as hardness, renitence, fixity, mobility, and condition of the subcutaneous plane are assessed [4]. The consistency of the swelling may suggest diagnostic hypotheses [7]. However,

it is virtually impossible to determine the clinical relationship between the tumor and the bone [7]. The differential diagnosis is made particularly with congenital cranial dysmorphism, such as trigonocephaly, plagiocephaly, or post-traumatic dysmorphia.

Conflict of Interest

The authors declare that they do not have any conflict of interest.

References

[1] Akhadar A, Gazzaz M, El Mostarchid B, Kadiri B, Lrhezzioui J, Boucetta M. Giant epidermoid cyst of the skull vault with extra- and intracranial development [Kyste épidermoïde géant de la voûte du crâne à développement extra et intra crânien]. *J Neuroradiol*. 2002;29:193–9.

[2] Akhadar A, Gazzaz M, El Mostarchid B, Kadiri B, Lrhezzioui J, Boucetta M. Giant epidermoid cyst of the skull vault with extra- and intracranial development [Kyste épidermoïde géant de la voûte du crâne à développement extra et intra crânien]. *J Neuroradiol*. 2002;29:193–9.

[3] Meder JF, Godon-Hardy S, Ricolfi F, Limot O, Aubert F, Lebtahi R, *et al.* Tumors of the cranial vault [Tumeurs de la voûte du crane]. *Feuillets Radiol*. 1993;33(1):16–31.

[4] Visot A, Civit T. Tumors of the vault and base of the skull. Part II, Intracranial tumors [Tumeurs de la voûte et de la base du crâne. Partie II, Tumeurs intracrâniennes] Neurochirurgie Ellipses edition, AUPELF/UREF, pp. 241–53.

[5] Jaiswal AK, Mahapatra AK. Giant intradiploic cysts of the skull: a report of eight cases. *Br J Neurosurg*. 2010;14(3):225–8.

[6] Sami A, Ben Ali S, El Kamar A, Boucetta M. Epidermoid cyst of the skull vault: a case report [Kyste épidermoïde de la voûte du crâne: a propos d'une observation]. *Neurochirurgie*. 1999;45(1): 58–61.

[7] Vinchon M, Lejeune JP, Krivosic I. Cranio-encephalic dermoid and epidermoid cysts: classification and pathogenesis [Les kystes dermoïdes et épidermoïdes cranio-encéphaliques: classification et pathogénie]. *Neurochirurgie*. 1995;41:29–37.

[8] David D. Anatomopathological and radiological correlation of cranial vault lesions and swellings in adults (Based on 32 cases) [Corrélation anatomo-pathologique et radiologique des lacunes et tuméfactions de la voûte crânienne chez l'adulte (A propos de 32 cas)]. Université de Picardie-UFR de médecine d'Amiens. 1985.

[9] Arana E, Latorre FF, Revert A, Menor F, Riesgo P, Liano F. Intradiploic epidermoid cysts. *Neuroradiology*. 1996;38:306–11.

[10] Patel TR, Borah GL. Frontal bone periosteal osteomas. *Plast Reconstr Surg*. 2004;114:648–51.

[11] Bhandari YS, Jones RA. Osteoma of the middle cranial fossa: case report. *J Neurosurg*. 1972;37:610–2.

[12] McClellan RL, Eisenberg RL, Givanni VL. Routine CT screening of psychiatric patients. *Radiology*. 1988;169:99–100.

[13] Dahlin DC. *Bone Tumors: General Aspects and Data on 6221 Cases*. 4th ed. Springfield: Charles C. Thomas; 1978.

[14] Schajowicz F. Bone-forming tumors. In *Tumors and Tumor-Like Lesions of Bone: Pathology, Radiology, and Treatment*. 2nd ed. Schajowicz F, Ed. Berlin: Springer-Verlag, 1994, pp. 29–140.

[15] Lee YY, van Tassel P, Nauert C, Raymond AK, Edeiken J. Craniofacial osteosarcomas: plain film, CT, and MR findings in 46 cases. *AJR Am J Roentgenol*. 1988;150:1397–402.

[16] Wilner D. *Radiology of Bone Tumors and Allied Disorders*. Philadelphia: Saunders; 1982, pp. 1897–2169.

[17] Rao VPK, Rout D, Radhakrishnan V. Osteogenic sarcoma of the skull. *Neuroradiology*. 1983;25:51–3.

[18] Salvati M, Ciappetta P, Raco A. Osteosarcomas of the skull: clinical remarks on 19 cases. *Cancer*. 1993;71(7):2210–6.

[19] Smeele LE, Kostense PJ, van der Waal I, Snow GB. Effect of chemotherapy on survival of craniofacial osteosarcoma: a systematic review of 201 patients. *J Clin Oncol*. 1997;15(1):363–7.

[20] Ben Salem B, Martin D, Cognet F, Beaurin J, Baudouin N, Couaillier J-F, *et al.* Diagnosis of skull vault lacunae in adults [Diagnostic des lacunes de la voûte du crâne chez l'adulte]. *RADIOLOGIE ET IMAGERIE MÉDICALE*. 1986;31645(A10):36–43.

[21] Rao VPK, Rout D, Radhakrishnan V. Osteogenic sarcoma of the skull. *Neuroradiology*. 1983;25:51–3.

[22] Schramek JK, Kassner EG, White SS. MR appearance of osteogenic sarcoma of the calvaria. *AJR Am J Roentgenol*. 1992;158:661–2.

[23] Chagnon S, Vallee C, Blery M, Chevrot A. Hemangioma and diffuse angiomatosis [Hémangiome et angiomatose diffuse]. *Encycl Med Chir (Paris). Radiodiagnostic Euroradiologie-Appareillocomoteur*. 1992;31510(A70):10–21.

[24] Tashiro T, Inoue Y, Nemoto Y, Mochizuki K, Katsuyama J, Hakuba A. Cavernous hemangioma of the clivus: case report and review of the literature. *AJNR Am J Neuroradiol*. 1991;12:1193–4.

[25] Steinbok P, Flodmark O, Norman MG, Chan KW, Fryer CJ. Primary Ewing's sarcoma of the base of the skull. *Neurosurgery*. 1986;19:104–7.

[26] Bhatoe HS, Deshpande GU. Primary cranial Ewing's sarcoma: short report. *Br J Surg*. 1998;12(2):165–9.

[27] Pritchard DJ, Dahlin DC, Dauphine RT, Taylor WF, Beabout JW. Ewing's sarcoma: a clinicopathological and statistical analysis of patients surviving five years or longer. *J Bone Joint Surg Am*. 1975;57:10–6.

[28] Desai KI, Nadkarni TD, Goel A, Muzumdar DP, Naresh KN, Nair CN. Ewing's sarcoma of the cranium. *Neurosurgery*. 2000;46(1):138–44.

[29] Kretzshmar HA, Eggert HR. Mesenchymal chondrosarcoma of the craniovertebral junction. *Clin Neurol Neurosurg*. 1990;92:343–7.

[30] Bose B. Primary osteogenic sarcoma of the skull. *Surg Neurol*. 2002;58:234–40.

[31] Ohnishi T, Oku Y, Yamamoto K, Morimura Y, Ushio Y. Benign chondroblastoma of the occipital bone. *Surg Neurol*. 1985;24:52–6.

[32] Artico M, De Caro GMF, Salvati M, Carloia S, Rastelli E, Wierzbicki V, *et al.* Solitary metastasis to the cranial vault: report of ten cases. *J Neurosurg Sci*. 2000;44:33–8.

[33] West MS, Russell EJ, Breit R, Sze G, Kim KS. Calvarial and skull base metastases: comparison of non-enhanced and Gd-DTPA-enhanced MR images. *Radiology*. 1990;174:85–91.

[34] Blackburn P. Visual diagnosis of hematologic and oncologic diseases. *Emerg Med Clin North Am*. 1993;11:273–5.

[35] Dias PS, May PL, Jakubowski J. Giant epidermoid cysts of the skull. *Br J Neurosurg*. 1989;3:51–8.

[36] White AK, Jenkins HA, Coker NJ. Intradiploic epidermoid cyst of the sphenoid wing. *Arch Otolaryngol Head Neck Surg*. 1987;113:995–9.

[37] Stark RJ, Henson R. Cerebral compression by myeloma. *J Neurol Neurosurg Psychiatry*. 1981;44:833–6.

[38] Resnick D, Kyriakos M, Greenway GD. Tumors and tumor-like lesions of the bone: Imaging and pathology of specific lesions. In *Bone and Joint Imaging*. Resnick D, Ed. Philadelphia: WB Saunders, 1989, pp. 1107–81.

[39] Khouja N, Aouidj L, Bahri K, Jemel H, Haouet S, Khaldi M. Solitary plasmacytoma of the skull vault: a case report and literature review [Plasmocytome solitaire de la voûte du crâne: a propos d'un cas et revue de la littérature]. *Neurochirurgie*. 2000;46(1):43–6.

[40] Bindal AK, Bindal RK, Van Lovern H, Sawaya R. Management of intracranial plasmocytoma. *J Neurosurg*. 1995;83:218–21.

[41] Braustein EM, White SJ. Non-Hodgkin lymphoma of bone. *Radiology*. 1980;135:59–63.

[42] Loembe PM, Minko-Mi Etoua D, MekeuzaM'Obiang L, Assengone-Zeh Y, Mwanyombet-Ompounga L. Primary lymphoma of the skull vault with fronto-parietal localization: a case [Lymphome primitif de la voûte du crâne de localisation fronto-pariétale: un cas]. *Neurochirurgie*. 1994;40:369–71.

[43] Rohringer M, Sutherland G, Louw D. Incidence and clinicopathological features of meningioma. *J Neurosurg*. 1971;89:665–72.

[44] Healy JF, Marshall WH, Brahme FJ, White F. CT of cranial metastases with skull involvement. *J Comput Assist Tomogr*. 1985;9:725–7.