Congenital Cervical Teratoma: About a Case

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ABSTRACT

Congenital cervical teratomas are benign but serious germinative cell tumors. The possibility of air compression structures and the risk of invasion of vascular and nervous noble elements are crucial prognostic factors in this kind of tumor. Prenatal diagnosis raises on ultrasound examination precisising locoregional consequences of the tumor and surgical possibilities. In the propitious cases, prenatal MRI examination is useful to precise tumor's limits and cerebral status of the foetus. At birth, coordinate management involving anaesthetists, pediatricians and specialized surgeons decrease morbidity and mortality. The recent observation of a case gave us the opportunity to make a literature review of this exceptional affection. It was a newborn female, aged 25 days allowed for anterior cervical mass evolving since the birth associated with moderate dyspnea. Cervical ultrasound coupled to CT scan had objectified a heterogeneous mass with cystic areas and calcifications. The excision was completed, and the histopathological study revealed a mature teratoma. The postoperative course was uneventful, with some food and phonatory transient disorders. Cosmetic outcome at 6 months was excellent. Cervical teratoma is a rare tumor, diagnosed mainly in the neonatal period, which requires a prenatal diagnosis and multidisciplinary early management. The prognosis depends mainly on the presence of neonatal respiratory distress and histological form. There are opportunities sudden worsening respiratory and malignant transformation that justify early surgical excision and prolonged follow-up.

Keywords: teratoma, neck, newborn, support.

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I. INTRODUCTION

Congenital cervical teratoma is a very rare germ tumor in newborns. Diagnosed primarily in the neonatal period, it presents etiopathogenic problems and therapeutic management problems that must be early and multidisciplinary. The anterior seat of this tumour, and its often large volume, causes in the majority of cases a compression of the airway. The prognosis depends on the presence of neonatal respiratory distress and histological form.

II. OBSERVATION

This is a 25-day-old female newborn born from a non-inbred marriage, the pregnancy was not properly followed and presumed to be completed, the delivery went without incident by low-born. At birth, this little girl weighed 3100 g, had a voluminous anterior cervical mass lateralized to the left

overflowing on the thorax, gradually increasing in volume, measuring 8 cm long axis, polylobed, firm consistency and fix in relation to the shallow and deep planes. She had moderate respiratory discomfort. After conditioning, intubation was not considered necessary. The rest of the somatic examination was without particularity.

Cervical ultrasound had objectified the presence of a large mass with irregular outlines of heterogeneous echostucture containing echogenic (tissues) and other anechogenic (fluids) areas and calcifications without doppler vascularization. Cervical-thoracic computed tomography (CT) (Fig. 1 and 2) confirmed the tissue and cystic components of the mass, which measured 38 mm 77mm 78 mm, and deviated cervical structures and compressed the trachea. At the thoracic stage, the mass sat at the level of the anterior mediastinum and spread over the upper and middle stages coming into contact with the descending aorta and the right pulmonary artery without any sign of locoregional invasion.

Fig. 1. Axial Cervical Computed Tomography after Contrast Media Injection: Large, well-limited latero-cervical mass left solido-cystic heterogeneous. Tissue component has moderate enhancement.

Fig. 2. Axial thoracic computed tomography after contrast media injection: large, limited anterior mass of heterogeneous solido-cystic tissue that exerts a mass effect on the trachea.

The little girl was operated, the surgical procedure consisted of a complete removal of this tumor mass, by an incision of Paul André. The dissection was easy because the tumour was well encapsulated (Fig. 3 and 4).

The anatomopathological examination of the operating room revealed a mature pluritissue teratoma with a minor immature component (grade 1).

The post-operative effects were simple, with some transient phonatory and eating disorders. The aesthetic result at 6 months was excellent.

Fig. 3. Aspect of the tumour that appears large and well encapsulated.

Fig. 4. aspect of the tumour that appears large and well encapsulated.

III. DISCUSSION

Congenital teratomas occur in 1/4000 births, accounting for 25–35% of all neonatal tumours [1]. The most common locations are sacrococcygeales (60%) and gonadal (22%) [2]-[3] while cervical location (3%) remains rare [4]. In the literature, about 300 cases of cervical-facial teratomas have been reported, the first being presented by Hess in 1856 [5].

A. Prenatal Diagnosis

Positive prenatal diagnosis is based on the presence of a cervical mass with solid and liquid components. The main circumstances of discovery are visualization of cervical mass and polyhydramnios [1], [6], [7]. In fact, the diagnosis usually leads to an interruption of pregnancy, given the large size of the tumor or the complications already caused [6]-[10]. Antenatal diagnosis allows planning of pregnancy and immediate multidisciplinary neonatal management. In contrast, despite a reassuring antenatal evaluation and a decision to continue pregnancy, the primary neonatal risk remains ventilatory urgency and difficult intubation conditions that may lead to a life-threatening prognosis [11].

B. Neonatal Diagnosis

At birth, they are tumors very often very voluminous, which deform the cervical reliefs. From the anterior seat, they can compress the cervical organs, mainly the trachea. Depending on the tumor volume, it will be proposed a low birth or a caesarean section.

Teratomas are most frequently isolated tumours. The four major congenital malformation reported in association with cervical teratoma are imperforated anus, chondrodystrophy, left ventricle hypoplasia with mitral narrowing and pulmonary hypoplasia [12].

Cervical ultrasound is often sufficient to make the diagnosis. Computed tomography (CT), and especially magnetic resonance imaging (MRI) with contrast injection, allow to specify the reports of the tumor, especially with the large vessels of the neck [5]-[13].

C. Differential Diagnostics

In front of a cervical mass, it is necessary to mention the lymphatic malformations, a lymphangioma, a congenital goître, a cyst of the thyreoglobose tract, a gill cyst, an arteriovenous malformation, a lipoma, a laryngocele. It will also be necessary to evoke the tumors of nerve origin: schwannomas, plexiform neurosis and neurofibromas that are found in neurofibromatoses, along the nerve paths.

The main differential diagnoses are cystic lymphangioma and hygroma [14].

D. Diagnosis of Malignancy

When tumor markers such as alpha-fetoprotein and β HCG (β human chorionic gonado-trophin) are abnormal, it affirms the presence of a malignant secreting component in the teratoma even though it is not found by anatomopathologist [15]. In the case we presented, the little girl had not benefited from these dosages.

E. Support

The management strategy for giant cervical teratomas differs: most authors opt for a cesarean section from the outset regardless of the size of the tumor from 36 SA [16]. For the control of the airways, several alternatives are conceivable from the least to the most deleterious: endotracheal intubation, cricothyrotomy, or tracheotomy if necessary, in case of failure of the previous ones. The use of nasofibroscope, or even rigid laryngoscope-bronchoscope in an emergency situation, can prevent tracheotomy.

Surgical management of these tumours must be early because of the risk of respiratory distress and degeneration [17].

In the absence of surgical treatment, mortality is 80%. Moreover, in this situation or in the case of incomplete removal, the risk of malignant degeneration would reach more than 90% of cases [18], [19].

The anatomopathological examination is the only examination that makes it possible to carry with certainty the diagnosis of teratoma. The extreme histological polymorphism of these tumours requires multiple samples and fine histological sections, so as not to ignore an immature or malignant hearth, the presence of which conditions the prognosis [20].

For a benign teratoma, the risk of recurrence is almost nil if the exesis has been complete.

When the tumour is encapsulated or pseudo-encapsulated and non-invasive, dissection from surrounding structures is easier, and the prognosis is excellent [21]. As a result, recurrences are rare and, if necessary, the patient will be able to benefit from a new procedure of excision [22], [23].

Some teams [18], [24] have taken charge of newborns carrying malignant cervico-facial teratomas with or without metastases. Due to the rarity of reported cases of malignant cervico-facial neonatal teratomas, there is no well-codified therapeutic approach.

F. Monitoring

Whether for benign teratomas or malignant teratomas, clinical surveillance is essential for the search for recurrence or metastasis. For this, the clinical examinations must be regular and close by the specialist and the pediatrician. MRI is the non-invasive, non-irradiating exam of choice, especially since there is no tumour marker to observe the recurrence of the tumour or the onset of metastasis [25]. The role of fetal Alfa serum protein monitoring has not been demonstrated for cervical facial teratomas [5].

IV. CONCLUSION

Cervical teratoma is a rare tumour, diagnosed primarily in the neonatal period, which requires antenatal diagnosis and multidisciplinary early management. Its prognosis depends essentially on the presence of neonatal respiratory distress and histological form. The risk of sudden respiratory aggravation and malignant transformation justifies early surgical removal and prolonged follow-up.

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