Endoscopic Resection of Pilocytic Astrocytoma of the Third Ventricle: A Case Report

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ABSTRACT

Pilocytic astrocytoma of the third ventricle is a very rare localization. It occurs mainly during the first two decades of life. The tumor is most often revealed by an HTIC syndrome. Brain imaging allows for establishment of a diagnostic hypothesis, which is confirmed only after an anatomopathological study.

We report the case of a six-year-old child with an astrocytoma of the third ventricle. He was admitted with intracranial hypertension. A gross total resection was performed using trans-frontal trans-ventricular endoscopy. Histological examination confirmed the diagnosis of pilocytic astrocytoma. Postoperative follow-up was straightforward, and the child made a full recovery.

To our knowledge, only a few reports of intraventricular pilocytic astrocytomas have been published and none have described endoscopic resection in the pediatric population which can be a good alternative option in this case.

Keywords: Endoscopy, Pediatric, Pilocytic astrocytoma, Third ventricle.

1. Introduction

Pilocytic astrocytomas are World Health Organization grade I tumors. They represent approximately 5%–6% of all gliomas and are most commonly diagnosed between the ages of 8 and 13 years [1]. Although these tumors are typically located in medial CNS structures, such as the cerebellum, thalamus, and hypothalamus, they rarely arise in the cerebral ventricle [2].

Intraventricular pilocytic astrocytomas are rare and represent 4% to 15.6% of all pilocytic astrocytomas and the third ventricle. Pilocytic astrocytomas are exceptional [3]. They are tumors of the central nervous system that are mainly detected during the first two decades of life [4]. In contrast to other low-grade astrocytomas, pilocytic astrocytomas are associated with the absence of a mutation in the isocitrate dehydrogenase (IDH) gene mutation. In some cases, KIAA1549/BRAF fusion is present. Clinical presentation is dominated by intracranial hypertension syndrome. Brain MRI allows the diagnosis, but confirmation is only possible in an anatomopathological study. The effective treatment remains the microsurgical complete removal of the tumor with an excellent prognosis. Third ventricle endoscopy is routinely used for some benign tumors such as colloid cysts or arachnoid cysts [5].

We report a histologically and molecularly confirmed case of third ventricle pilocytic astrocytoma in a child who underwent complete endoscopic resection.

2. Case Report

Our patient was a six-year-old male child with no previous pathological history, presenting with HTIC syndrome with headache and vomiting that appeared four days before his consultation without any other associated signs. Clinical examination revealed a conscious patient with a perfectly normal neurological examination. Brain MRI showed a solid cystic tumor at the floor of the third ventricle of about 26 mm with low enhancement after injection of contrast medium with upstream tri-ventricular hydrocephalus without evidence of trans ependymal resorption [Fig. 1]. The histological type could not be specified. The lesion was approached endoscopically through a right ventricle. Intraoperatively, the tumor appeared cystic and well-limited [Fig. 2]. The resection was gross total, sparing the roof of the third ventricle, which was inflamed. The tumor was poorly vascularized with a little bleeding which was easily controllable by coagulation. The child recovered completely without any postoperative complications. Pathological examination showed a low cell density on a microcystic background loaded with Rosenthal fibers. The
Fig. 1. Axial: A and sagittal; B injected T1 MRI revealing the process on the floor of the third ventricle.

Fig. 2. Endoscopic view showing the intraoperative solido cystic aspect of the process.

cells were piloid in appearance with a tapered nucleus and the diagnosis of pilocytic astrocytoma was retained.

3. Discussion

Pilocytic astrocytoma is the most common central nervous system (CNS) tumor in the pediatric population and accounts for approximately 5%–6% of all gliomas [5]. It represents 1.3% of all CNS tumors and one-third of patients aged 0–14 years [1], [6]. It can occur anywhere in the cerebral nervous system, although it most commonly occurs in the cerebellum (42%), followed by the supratentorial compartment (36%), optic tract and hypothalamus (9%), brainstem (9%), and spinal cord (2%). In children, the most frequently affected site is the cerebellum (67%), with only rare cases developing supratentorial [7]. In exceptional cases, they occur in other locations, such as the intraventricular space [1].

Intraventricular neoplasms arise from cells forming the ependymal mucosa or subependymal plate of the ventricular wall, choroid plexuses, and glia-lined structures such as the septum pellucidum [8]. Intraventricular tumors are relatively asymptomatic until they enlarge and obstruct the CSF pathways, producing obstructive hydrocephalus and leading to increased intracranial pressure. The typical appearance of pilocytic astrocytoma on magnetic resonance imaging and computed tomography is that of a cyst with an intensely enhanced mural nodule [9]. However, the radiological features of intraventricular and extra ventricular pilocytic astrocytomas were similar to typical features, including the enhanced nodules in cysts. Radiological findings are generally diagnostic of this tumor entity [10]. Histologically the classic pilocytic astrocytoma shows a biphasic growth pattern including compact cellular areas and loose, microcystic areas with Rosenthal fibers and eosinophilic granular bodies. Immunohistochemical studies have shown that tumor cells are positive for glial fibrillary acid protein (GFAP), ATRX, and Olig2 with a Ki67 labeling index of less than 1%, indicating the astrocytic nature of the tumor. Molecular studies revealed that there is no mutation of the IDH gene [11].

Surgical management in the majority of reported cases is classical resection using microsurgery a trans-lateral ventricular trans-foramen of Monroi approach. Endoscopic surgery is very interesting for the approach of the third ventricle even in the case of a young child. In our case, a small frontal incision was sufficient to visualize the cystic tumor, remove it, and free the monro hole which was clearly visible. Endoscopy allowed to optimize the management by avoiding the use of the microscope which only allows to zoom the operative field and not to navigate in the ventricular system. At the same time, this therapeutic choice is more aesthetic for the child and less disabling. Total resection is the best management choice and has a low risk of tumor recurrence [12]. But if the tumor infiltrates the walls of the third ventricle, we can leave the tumor which was adherent to the floor of the third ventricle to minimize the hypothalamus injury. Pilocytic astrocytoma generally has an excellent prognosis. Total excision. In case of tumor residue, close monitoring should be performed in order to initiate adjuvant therapy or surgical revision.

4. Conclusion

Pilocytic astrocytomias of the third ventricle are very rare tumors that are difficult to diagnose preoperatively on the basis of radiological profile alone. The most frequent manifestation is hydrocephalus, so it is essential to restore normal pressure dynamics in the intracranial space. The endoscopic approach remains the best therapeutic choice, which we strongly recommend, for tumor exeresis and freeing the cerebrospinal fluid outflow tracts. The prognosis remains excellent in case of total removal of the tumor.

Conflict of Interest

Authors declare that they do not have any conflict of interest.

References


