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Pilocytic Astrocytoma in the Sellar and Suprasellar Region Mimicking Craniopharyngioma: A Case Report

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Abstract **Case Report**

***Introduction*:** Pilocytic astrocytoma, represents 5-6% of glial tumours, classified as grade I by the World Health Organisation. It tends to grow slowly and often contains cysts. ***Case report*:** We report the case of a young girl aged 6 years and 5 months, presenting a huge heterogeneous intra and supra sellar mass (87*65*44 mm) suggesting a craniopharyngioma. Anatomopathological and immunohistochemical examination revealed a tumoral proliferation compatible with a grade 1 pilocytic astrocytoma. A*djuvant chemotherapy* and radiotherapy were indicated. ***Discussion & Conclusion*:** The diagnosis of pilocytic astrocytoma is suggested after magnetic resonance imaging which specifies the topography, size, infiltrative or expansive nature of the lesions. However the sellar/suprasellar location of the pilocytic astrocytoma contribute significantly to the diagnostic difficulty. The histological examination allows the diagnosis to be confirmed after biopsy or resection of the tumour and to consider the strategy for further treatment. Management of sellar and suprasellar pilocytic astrocytoma remains challenging and requires a multidisciplinary approach.

Keywords: Pilocytic astrocytoma - Sellar/Suprasellar - Management - Transsphenoidal surgery.

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INTRODUCTION

Pilocytic astrocytoma (PA), previously known as cystic cerebellar astrocytoma or juvenile pilocytic astrocytoma, is a grade I glial neoplasm, represents 5-6% of glial tumours, It tends to grow slowly and often contains cysts [1]. It usually develops during the first two decades of life and it represents the most common cerebral tumour (18%) in the paediatric age [2]. It's generally a well-circumscribed, often cystic astrocytoma composed of variable proportions of loose and compact tissue [1]. Clinical symptoms of PA vary depending on the tumor size, location and extension, and age at diagnosis [3]. They typically arise in deep, midline structures such as the cerebellum, brain stem, optic nerve and infundibulum, whereas lobar tumours are rare [4]**,** Unusual intracranial locations of PA have been reported to occur, including sellar areas [5].

We present our experience with a patient with intrasellar and suprasellar pilocytic astrocytoma mimicking craniopharyngioma removed through the transsphenoidal approach.

CASE REPORT

We report the case of a young girl aged 6 years and 5 months, who consulted for a decreased visual acuity, convergent strabismus of the right eye, and intermittent headaches without vomiting. The clinical examination noted a blood pressure of 108/78 mmHg, heart rate of 78 bpm, Weight of 41kg (+3 SDS), Height of 127cm (+1 SDS), bilateral nystagmus. Brain MRI (Figure 1) showed a huge heterogeneous intra and supra sellar mass (87*65*44 mm) extended to the cavernous sinus with compression of the carotid artery and extension through the left optic hole suggesting a craniopharyngioma. Initial monitoring of pituitary functions noted corticotropic deficit substituted with hydrocortisone. The patient underwent surgical resection *via* a *transsphenoidal approach.* Postoperative course was simple, she presented diabetes insipidus treated with Desmopressin, also the hypophysogram showed the pre-existing corticotropic lack without other pituitary deficits. Anatomopathological and immunohistochemical examination revealed a tumoral proliferation compatible with a grade 1 pilocytic astrocytoma. A*djuvant chemotherapy* and radiotherapy

were indicated. Postoperative visual field examination showed right temporal hemianopia, and only a small spot in the superior temporal quadrant is perceived in the left eye. Continuous monitoring of hypothalamic-pituitary functions, during the follow-up, is necessary for the early diagnosis of other endocrine disorders.

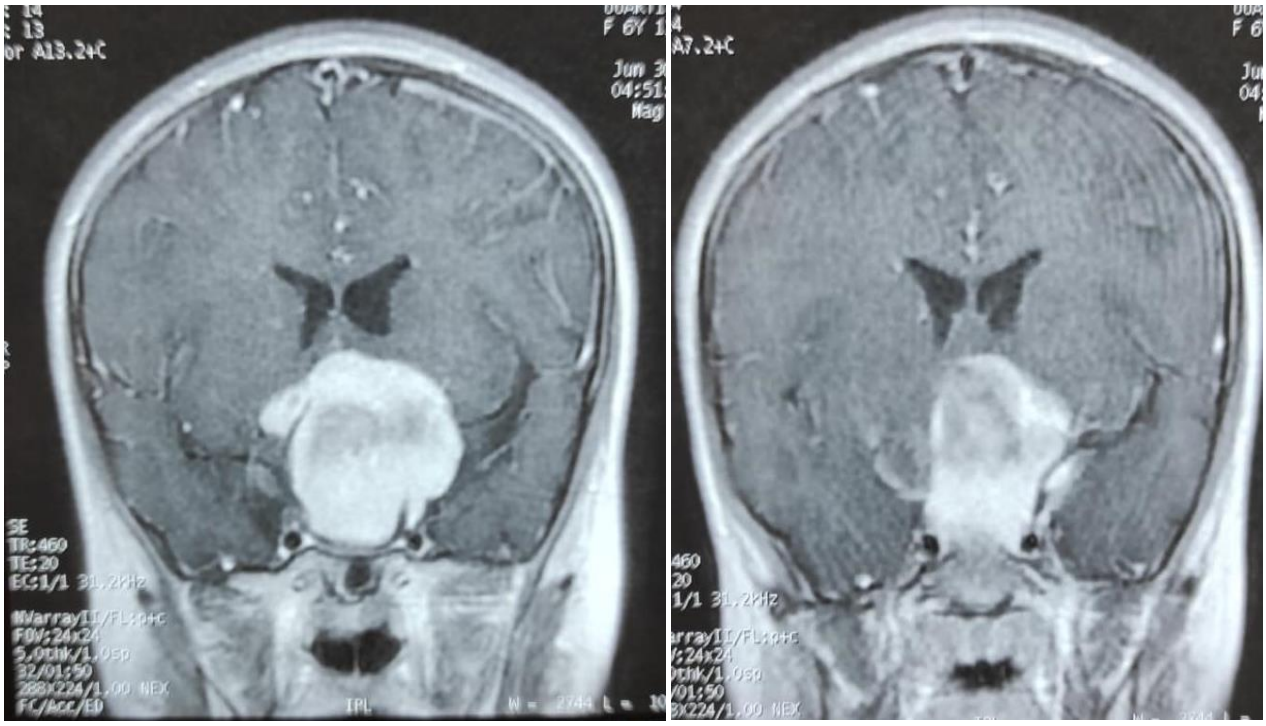

Figure: Coronal MRI images illustrating a large sellar and suprasellar heterogenous mass with the radiological appearances of a craniopharyngioma

DISCUSSION

Pilocytic astrocytoma (PA) is a rare, slow-growing glioma, classified as grade I by the World Health Organisation; it's the most common glial neoplasm in children [6].

PA represents 6% of all primary intracranial tumors, rarely found in the pituitary fossa. It is not usually considered in the differential diagnosis of an intrasellar tumor, as the most common neoplasms of the pituitary fossa are pituitary adenomas, granular cell tumors, craniopharyngiomas, chordomas germ, gliomas, meningiomas... [5]

In pediatric patients, two-thirds of lesions are located in the cerebellum; however, the entire neuraxis can be involved. There are no clinical characteristics that are pathognomonic to these tumours. Signs and symptoms are determined by tumoral size, location and the presence or absence of hydrocephalus [6]. PA arising in sellar and suprasellar regions may cause hypothalamic-pituitary dysfunction, and visual loss or visual-field deficit if optic pathway is involved [1, 2].

In our case, the sellar/suprasellar site of the PA contributed significantly to the diagnostic difficulty. It had clinical and radiological features compatibe with diagnosis of craniopharyngioma and was only confirmed to be pilocytic astrocytoma after histopathological examination.

Standard primary management of PA in both children and adults consists on maximal safe surgical resection [7-9]. Sellar neoplasms can be removed by transsphenoidal approach with minimal morbidity [10]. Cerebellum involvement allows total resection, however, when tumors are located at other sites (Chiasmatic-hypothalamic or midline tumors ...), or when there is residual tumor, less favorable results may be obtained [8, 11].

Management after surgical resection of the tumor is less clear and depends on extent of resection [7]. If removal is complete, no adjuvant therapy is necessary, Observation is recommended. A second or even third surgical resection is often required in children with progressive/recurrent disease, when a complete or near-complete resection can also be achieved. For PA located in critical or deep areas (such as the brainstem and hypothalamus...) complete resection in usually not achievable [6-8].

In cases of incomplete resection, the need for immediate additional therapy such as chemotherapy or radiotherapy remains controversial among clinicians. Chemotherapy is used preferably in young children to avoid the toxicity of radiotherapy. However, there is no consensus on when radiotherapy should be used in

children, or at what age radiotherapy should be used rather than chemotherapy [7, 8].

PA is the glioma with the most favorable prognosis, the success of its treatment therefore depends on the location of the tumor and the quality of surgical intervention [11]. Complete resection has been associated with improved progression-free and overall survival. 10-year overall survival rates of 90-95.8% were reported after total resection [4, 9]. Chiasmatic-hypothalamic or midline tumors, in which complete resection is more difficult to perform, have shown poorer progression-free survival than cerebellar or cerebral tumors [8, 9]. In case of subtotal resection, 20-year survival rates of 70 to 80% were observed [12].

CONCLUSION

In conclusion, due to its rarity, there's few data regarding management and prognosis of pilocytic astrocytomas arising in the sellar/suprasellar region. First line therapy and the main therapeutic goal is to achieve complete surgical resection, however the treatment of this entity, particularly when it involves structures such as hypothalamus, pituitary gland and optic pathway remains challenging and requires a multidisciplinary approach.

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