

Pilocytic astrocytoma, another way to reach your diagnosis

Astrocitoma pilocítico, outra forma de chegar ao seu diagnóstico

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ABSTRACT

Cerebellar astrocytoma (low-grade glioma) is the most frequent tumor of the Central Nervous System in pediatric age, corresponding to 10-20% of brain tumors, having its maximum incidence at 5 years. Brain tumors are the second cause of death at this age, behind leukemias. Its most frequent clinic is headache with vomiting which can worsen in the morning and awaken the patient at night. The most frequent ophthalmological clinic is papilledema and involvement of the cranial nerve VI. In our case we present an atypical presentation (cranial IV), in which a quick derivation favored a better prognosis.

Keywords: Astrocytoma; Glioma; Intracranial hypertension; Brain Neoplasms; Child

RESUMO

O astrocitoma cerebelar (glioma de baixo grau) é o tumor mais frequente do Sistema Nervoso Central em idade pediátrica, correspondendo a 10-20% dos tumores cerebrais, tendo sua incidência máxima em 5 anos. Os tumores cerebrais são a segunda causa de morte nesta idade, atrás das leucemias. Sua clínica mais frequente é a cefaleia com vômitos que podem piorar pela manhã e despertar o paciente à noite. A clínica oftalmológica mais frequente é o papiledema e o envolvimento do nervo craniano VI. Em nosso caso apresentamos uma apresentação atípica (IV craniana), em que uma derivação rápida favoreceu um melhor prognóstico.

Descritores: Astrocitoma; Glioma; Hipertensão Intracraniana; Neoplasias encefálicas; Criança

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INTRODUCTION

Cerebellar astrocytoma (low-grade glioma) is the most frequent tumor of the central nervous system in pediatric age, corresponding to 10-20% of brain tumors, having its maximum incidence at 5 years. Brain tumours are the second cause of death at this age, behind leukaemias.⁽¹⁾

Clinical Case

A 4 year old patient who comes to an ophthalmology consultation because the mother refers that her daughter takes strange positions to read. Intense headache of morning predominance, without associated vomiting, waking her up some nights. No alterations of speech or stability. No loss of visual acuity in any eye. The patient's head is rotated to the right and inclined over the left shoulder. No restriction in the mobility of the clear eyes due to bad collaboration. Slight hypertropy in primary right eye position [suspected right IV cranial nerve (cn)]. No involvement of more cn. Cover/uncover negative. Anterior segment without alterations.

In funduscopy, clear bilateral papilla edema with erasure of borders, congestive signs, without peripapillary haemorrhages is observed (Figure 1). The rest of the fundus of the eye is normal, presenting neither hemorrhages in the retina nor alterations in the macula. Given the suspicion of intracranial hypertension with possible involvement of the right IV cn (due to compression of the roof of the IV ventricle, affecting the posterior aspect of the midbrain, where the cranial nerve exits), Computerized Axial Tomography (CAT) is performed (Figure 2A).

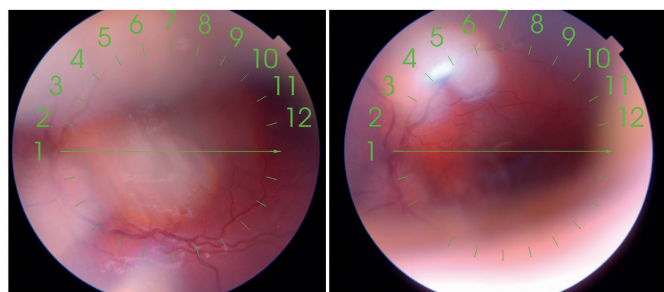


Figure 1: Papilla edema

DISCUSSION

Brain tumors are the second leading cause of tumors in children under age 15. The annual incidence is 2-5 cases/100,000 per year, with greater involvement of males. There is no difference by race. The 54% supratentorial, 40% are infratentorial, and 6% medullary. Diagnosis is made by imaging tests and treatment is usually surgical adjuvant with oncological treatment.^(2,3)

In adults, the most frequent brain tumors are brain metastases, while the most frequent primary is glioblastoma multiforme. However, in children, the most common tumour is pilocytic astrocytoma (low-grade glioma) and the most common malignant tumour is medulloblastoma. Metastases are very rare. The symptoms depend mainly on the location but the most frequent is headache. It is usually more intense in the morning and may wake patients at night.

Gliomas are heterogeneous group of tumours that are classified histologically according to the criteria of the World Health Organisation (WHO) in 2 groups, low and high grade. The low grade are divided into pilocytic astrocytoma (grade 1 tumors) or fibrillary astrocytoma (grade 2).^(4,5)

Pilocytic astrocytoma is the most common brain tumour

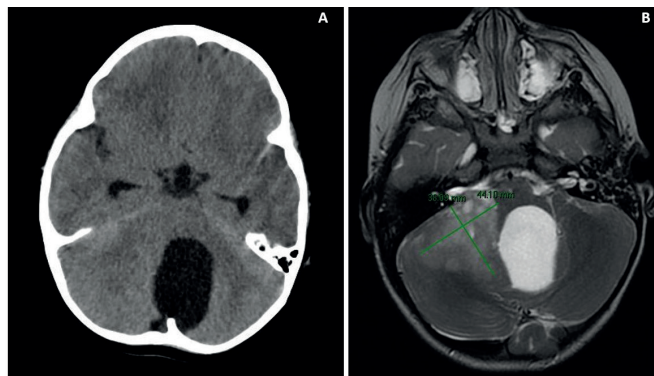


Figure 2: A: CT scan prior to surgery. B: NMR with contrast prior to surgery.

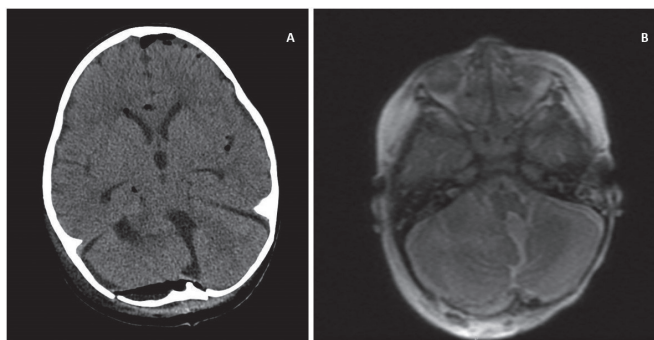


Figure 3: A. CT after surgery. B. NMR without contrast after surgery

in children, they are slow-growing tumours with little risk of metastasis, they act mainly by displacing tissues. The prognosis of astrocytomas will depend mainly on the histological type, age of diagnosis, size and location of the tumour.⁽⁶⁾

Due to intracranial hypertension the cranial nerve most affected the VI pc and the most frequent alteration in the fundus of the eye the papilledema. In our case, the study was initiated due to the discovery of papilledema and the affectionation of the right IV cn.

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