Metastatic Neuroblastoma with Initial Orbital Presentation

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ABSTRACT

Neuroblastoma, also called sympathoblastoma, is a malignant embryonal tumor developed at the expense of neural crest cells, the most frequent solid tumor in children under 5 years of age.

The objective is to recall the clinical variability and the importance of evoking neuroblastoma in a child presenting with an ophthalmological symptom.

We report the case of a 5-year-old child who presented with a right orbital metastatic neuroblastoma, whose ophthalmologic complaint led to the diagnosis.

An ophthalmologist in the emergency room for a V AD and eye pain first saw the child. On examination, true exophthalmos with palpebral edema was found on the right. Cerebral and orbital CT revealed a right latero-orbital osteolytic mass of secondary appearance with endocranial extension. The workup showed lymph node involvement, unilateral right adrenal, osteomedullary, hepatic, and pancreatic involvement.

The child was admitted to a specialized pediatric oncology department and received chemotherapy. The patient was then declared under palliative treatment.

Keywords: Exophthalmos, metastasis, Neuroblastoma.

1. Introduction

1.1. Observation

Neuroblastoma, also called sympathoblastoma, is a malignant embryonal tumor developed at the expense of neural crest cells. It can develop anywhere in the body where sympathetic nerve cells exist. It is the most frequent malignant tumor of solid tumors in children under 5 years old. Orbital metastasis may be indicative of the primary tumor (10% of cases). Primary orbital involvement is exceptional [1].

2. Clinical Case

2.1. Observation

We report the case of a 4-and-a-half-year-old child, the youngest of two siblings, born to healthy non-consanguineous parents, with no particular medical, surgical, or ophthalmological history, well vaccinated according to the national immunization program.

Consulted for ocular pain with a decrease in visual acuity dating back 10 days without any traumatic context.

Right eye: Visual acuity is difficult to quantify. Palpebral edema; exophthalmos of the right eye (Fig. 1); corneal ulcer with positive fluorescence (Fig. 2).

Left eye: Visual acuity difficult to evaluate; oculomotor reflexes preserved; anterior and posterior segments without any particularities.

Fig. 1. Image showing Exophthalmos of the right eye.
A cerebral and orbital CT scan was ordered and revealed a right latero-orbital osteolytic mass of secondary appearance with endocranial extension (Fig. 3).

The patient was subsequently referred to a pediatric oncology center for further diagnosis and management.

The patient underwent primary chemotherapy for neuroblastoma and the extension workup showed lymph node involvement, unilateral right adrenal, osteomedullary, hepatic and pancreatic.

The child was declared under palliative treatment.

3. Discussion

Neuroblastomas are malignant neuroblastic tumors of the sympathetic nervous system and are, as well as the corresponding benign tumors - the ganglioneuromas -, developed from neural crest cells [2]. Orbital neuroblastoma in children commonly occurs due to metastasis with primary in the abdomen [3]. Forty percent of orbital metastases are bilateral [3].

The diagnosis of neuroblastoma is often delayed due to its varying and nonspecific presenting symptoms [4].

Several review articles have summarized the various ocular manifestations of neuroblastoma in children [5]–[7]. Exophthalmos with lid and orbital ecchymoses and an abdominal mass are found in approximately 20% of all cases. Supraorbital masses, dilated scalp veins, and adnexal edema are frequent. Hydrocephalus, papilledema, dilated retinal vessels, pareses of extraocular muscles, ptosis, and ischemic optic neuropathy are reported less frequently.

4. Conclusion

Neuroblastoma is an aggressive tumor of the child whose remission can be obtained in particular thanks to an early diagnosis; one should not neglect the orbital metastatic presentation, which could be the initial sign of the disease.
CONFLICT OF INTEREST

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

REFERENCES


