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Intraorbital Mature Teratoma in Infant

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Abstract

Intraorbital mature teratoma is a rare congenital tumor of slow evolution that can compromise vision. **Objectives:** We describe the clinical and therapeutic aspects of a mature teratoma in childhood with a literature review. **Clinical Case:** It was a 12-year-old who had a progressive straight monocular exophthalmia with blindness. CT without and with contrast injection showed a double component intracolonic invasive tumor with partial contrast acquisition after injection. This tumor respected the bone walls of the orbit. The treatment was surgical by conjunctival dissection of the tumor and enucleation. This allowed the tumor to be fully excised under an operating microscope. The operating procedures were simple. Anatomopathology confirmed the nature of the tumor by visualizing the presence of a squamous epithelium by the high-magnification (X20) method of hematoxylin and eosin. **Conclusion:** Mature intraorbital teratoma is an embryonic tumor with early diagnosis the finding in an older child remains rare. Complete surgical excision allows healing.

Keywords

Teratoma, Mature, Micro Surgery

1. Introduction

The mature intra-orbital teratoma is a rare tumor and represents 1% of the orbital tumors [1]. From embryonic origin, it is due to abnormal cell development between the 3rd and 5th week of embryonic life [1]. The origin of teratoma has been derived from the Greek word "Teratos" and "Oma". The literal meaning of which accounts to teratos meaning unusual occurrence or phenomenon, monster

and oma meaning complete set of, or a condition [2]. A slow evolution can be discovered in the older child [3]. Visual impairment includes major visual impairment and ophthalmoplegia. We present a rare case of right monocular blindness revealed by grade 3 exophthalmia and discuss the diagnostic and therapeutic features.

2. Clinical Case

This was a 12-year-old girl who was seen in consultation for exophthalmia and blindness of the right eye. The interrogation found a long history of eye pain and a progressive eye protrusion that has evolved over the past 6 years. The clinical examination of the eye revealed a grade 3 exophthalmia with the upper and lower eyelids separated, resulting in deformation of the face. It was also noted keratinization of the pupil and edematous infiltration of the white eye and conjunctival mucosa (**Figure 1**). There was no light perception or corneal reflex. Ophthalmological examination confirmed the right monocular blindness. The CT scan revealed a hypodense tissue expansive process of the orbital cavity pushing the eyeball from inside out (**Figure 2**). The bone and cartilage walls are pushed back, apart without erosions or osteolysis on the bone window sections. After injection of the contrast product, there was a partial hyperdense enhancement.

The surgical procedure consisted of enucleation by conjunctival dissection under an operating microscope. The tumor was resected with the eyeball and the optic nerve, which revealed a pearly white substance flowing under pressure (**Figure 3**) and another tissue of firm yellowish slightly calcified consistency. The operating sequences were simply marked by intraorbital healing with the local dressing care (**Figure 4**). She was reviewed for 3 months and 6 months after the surgery.

The anatomopathology examination found a squamous epithelium (**Figure 5**) after hematoxylin and eosin staining compatible with a mature teratoma.

3. Discussion

Teratomas are embryonic tumors derived from the ectoderm, endoderm and mesoderm [4]. It is a benign tumor and the sites of appearance are, in order of frequency, the ovarian, testicular, mediastinal anterior, retroperitoneum, precarious and coccygeal regions [5] Orbital location remains rare. A slight predominance of the left orbit was reported and the female sex was also mainly affected [6] but the tumor was located in the right eye in our case. This tumor, although benign, evolves in intraconic by progressively assaulting the eyeball responsible for exophthalmia and major visual disorders [7]. The persistent enlargement of this neoplasm is attributed to mucus secretion from the embryonic intestinal tissue [8].

Other features include; no family history of congenital deformities with non-consanguineous parents and normal siblings, normal pregnancy and delivery, and no history of teratogenic influences on the mother [9].

Figure 1. Front photo showing exophthalmos.

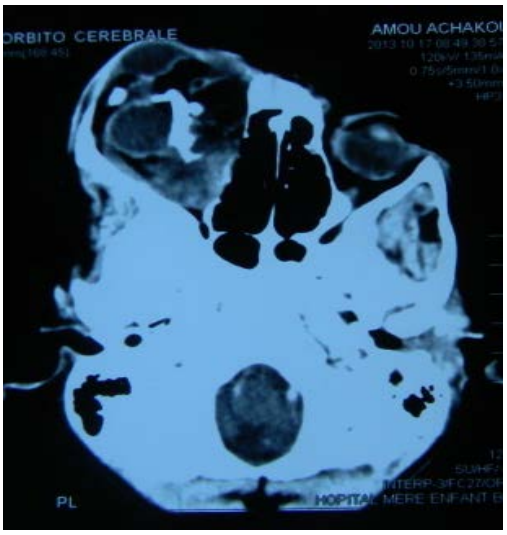

Figure 2. CTscan visualizing the orbital tumor with contrast enhancement.

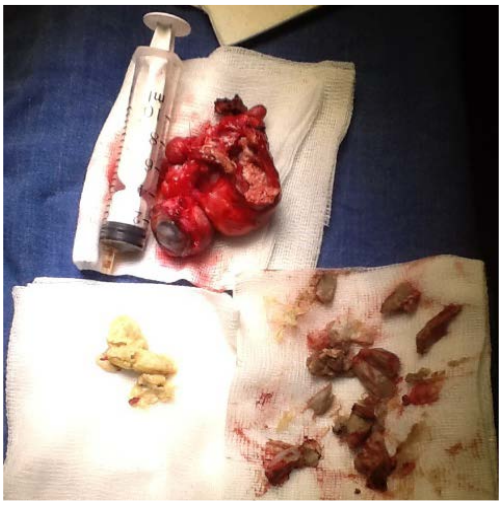

Figure 3. Enucleation image with tumor excision.

Figure 4. Postoperative photograph showing the right orbital cavity.

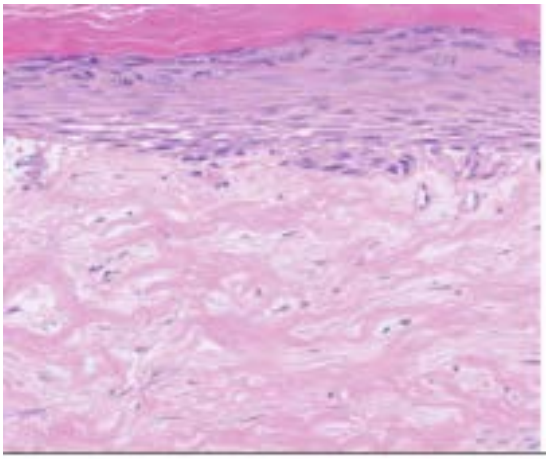

Figure 5. Photograph of the mature teratoma showing the squamous epithelium.

As in our patient, the tumor was responsible for right monocular blindness with exophthalmia. Computed tomography and magnetic resonance imaging make the diagnosis. The tumor remains intraconic without reaching the bone walls with mixed density [7] [10]. Enucleation excludes the tumor and reveals the presence of a squamous epithelium after hematoxylin and eosin staining [10]. In the case of mature teratoma, the complete exeresis gives good prognosis, as in our case if there are persistent immature cells [3], complementary radiotherapy and chemotherapy are recommended. Early surgery is mandatory to avoid permanent sequelae. The treatment is complete tumor excision with sparing of the eye, if possible. It is hard to recommend a fixed management plan, however, a common agreement in the management objectives should be to save the eye, retain some vision, encourage normal orbitofacial development, and maintain good cosmetic result [9].

4. Conclusions

The intraorbital mature teratoma is a rare embryonic tumor capable of compromising vision. Complete exeresis allows healing. Confirmation diagnosis is pathological by identifying a squamous epithelium with hematoxylin and eosin

after high magnification.

We obtained the consent of the patient and her family to publish these photographs.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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