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Case Report

Orbital Rhabfomyosarcoma in the Child: About Case

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Introduction

Rhabdomyosarcoma is a rare tumor, representing 5% of tumors occurring in children [1]. Its location is orbital in 10% of cases [1] and eyelid localization is exceptional [2]. It is an extremely virulent malignant tumor whose early diagnosis considerably improves survival and visual prognosis [3]. We report the case of a right orbital rhabdomyosarcoma presenting as isolated right exophthalmos of evolution in a 13-year-old child.

Observation

We report the observation of a 13-year-old child, without notable pathological history, who has presented for one month with progressive right exophthalmos. The rest of the somatic examination is unremarkable. A cervicofacial CT scan in favor of a hyperdense and heterogeneous intraconical tumor-like process above and retro-ocular right eye after injection of PDC, measuring 31*29 mm in height, this process is in intimate contact with the postero-internal part of the eyeball, without fatty border of separation and without visible endo-ocular bud, it is responsible for a repression of the eyeball with grade 2 exophthalmos (Figure 1). It encompasses and stretches the superior rectus muscle and the other rectus muscles as well as the optic nerve are respected. No lytic bone lesion or suspicious contrast enhancement was visualized at the cerebral and meningeal levels. An extension assessment made by a thoraco-abdominal-pelvic CT scan was normal. A biopsy was performed and the histological and immunohistochemical examination was in favor of a type C alveolar rhabdomyosarcoma (Figure 2). Given the size of the tumor and the local functional risk, surgery was rejected and chemotherapy was started according to the RMS 2005 protocol, which consists of 5 courses combining ifosfamide, vincristine and actinomycin (IVA). A treatment response assessment was carried out after three courses: the orbital CT scan revealed a clear decrease in size of the right intra- and extra-orbital tumor process, centered on the superior wall of the orbit and developed at the expense of the superior rectus muscle, with tissue density, enhanced after contrast and measuring 29*23*13 mm versus 54*55*54 mm. Given the persistence of this tumor residue: 3 courses of IVA were added, an orbital MRI for evaluation was done after the 8th course of IVA and was in favor of a very small residue centered on the superior rectus muscle of the right orbit without mass syndrome or pathological contrast uptake seen only on two sections of the control CT scan (Figure 3).

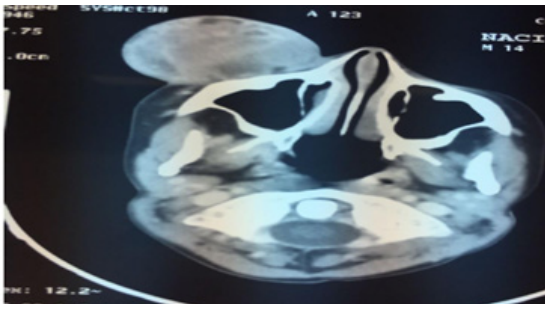

Figure 1: Axial and frontal CT image of an intra- and extra-conical orbital process pushing the right globe forward, causing exophthalmos.

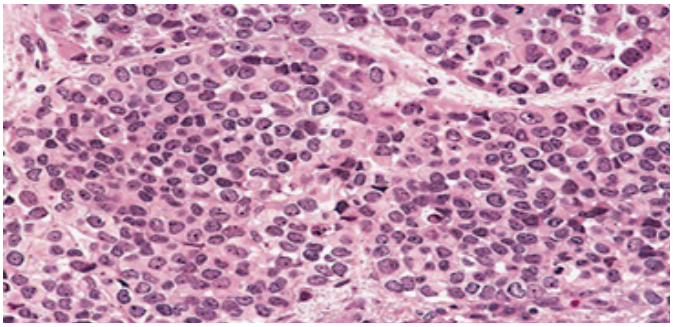

Figure 2: Histological examination, magnification 640x451.

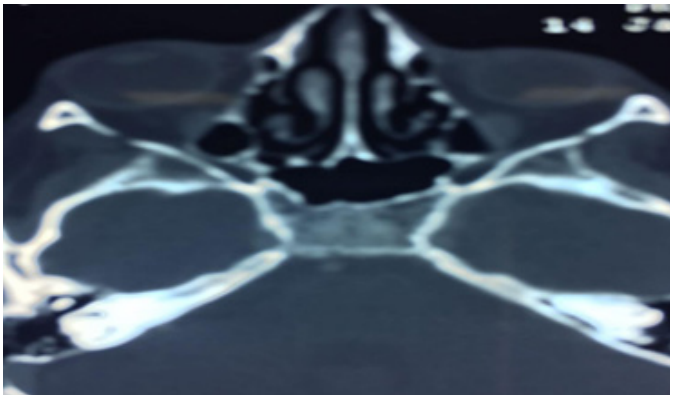

Figure 3: Axial section of an orbital CT scan after response to chemotherapy with persistence of a small remnant centered on the superior rectus muscle.

Intensity-modulated external beam radiotherapy (IMRT) was indicated after chemotherapy, on the tumor residue, in this patient at a total dose of 50 Gy: a classic fractionation 2Gy/fr in 25 sessions, with good clinical tolerance to external beam radiotherapy. An orbital MRI performed 6 months after external beam radiotherapy showed a small post-radiation inflammatory thickening of the superior rectus muscle, without pathological contrast uptake.

Discussion

Rhabdomyosarcoma is a rare tumor (5% of childhood tumors) with the most frequent orbital location (10% of cases) [1]. This tumor, which mimics striated muscle tissue and develops from embryonic mesenchymal cells, can arise both in striated muscle and in an area devoid of it [5,6]. Its preferred locations are, in order of frequency, the head and neck (40%), the genitourinary system (20%), the extremities (20%), and the trunk (10%) [7]. Among orbital locations, 76% are located in the orbital cavity; 12% are conjunctival, 9% intraocular, and only 3% are palpebral [2], which distinguishes this case from the usual presentations. Intraocular localizations are exceptional and develop from the ciliary body or the iris [8-11]. From a radiological point of view, imaging is not pathognomonic, but provides arguments in favor of the diagnosis [14]: tissue density of the lesion, enhancement after injection of iodine or gadolinium, aspects of osteolysis of the orbital walls. It allows the lesion to be visualized, to be located in the orbit, sometimes to confirm its original structure, to measure it, to identify its relationships and to specify its orbital or encephalic extension. This observation illustrates, by the normality of the initial imaging, the very rapid tumor growth [15,16] of rhabdomyosarcoma which makes it an emergency. The different histological forms of rhabdomyosarcoma are embryonic, alveolar, pleomorphic which are rarely localized to the orbit [2]. The contribution of immunohistochemistry is valuable in this type of tumor and allows certain erroneous diagnoses to be corrected. Indeed, the distinction between rhabdomyosarcoma and other mesenchymal tumors is sometimes difficult, especially when it comes to undifferentiated forms, as in this observation [17]. Management, which may include chemotherapy, surgery, and/or radiotherapy, requires multidisciplinary consultation. Surgery is not routine. The Intergroup Rhabdomyosarcoma Study Group I (IRSG I) prioritizes preservation of function over complete resection [3]. Thus, due to the initial tumor volume and to avoid mutilating excision, it was decided not to resort to surgery in this child. Rhabdomyosarcoma is a chemo-sensitive tumor. Current protocols combine three molecules when the tumor is not metastatic: vincristine, actinomycin, and cyclophosphamide; vincristine, actinomycin, and ifosfamide; vincristine, etoposide, and ifosfamide. Superior efficacy of the vincristine and actinomycin combination has been demonstrated [18]. Rhabdomyosarcoma is also a radiosensitive tumor, but requires high doses that are not without adverse effects. The main adverse effects are radiation cataract (55%), dry eye (36%), orbital hypoplasia (24%), ptosis (9%), and radiation retinopathy (90%) [12]. In this child, irradiation was only undertaken when imaging confirmed the existence of tumor residue after 6 chemotherapy sessions. Indeed, this technique does not affect patient survival [19] and protects them from a certain number of adverse effects of radiotherapy. Since the introduction of multimodal treatment by chemotherapy, surgery and/or radiotherapy, the survival rate of patients with rhabdomyosarcomas has significantly improved. Locally, regression is observed in 80% of cases (20% recurrences). Regionally, no lymph node spread is observed in 94% of cases; 6% of cases develop lymphadenopathy. Finally, generally, 94% of patients do not present systemic spread compared to 6% of cases where visceral metastases appear. Overall, 5-year survival is 94% for the embryonic form and 74% for the alveolar form [15]. Survival depends in particular on the non-metastatic nature of the disease. Added to this is the local functional risk, in particular of amblyopia and compression of the optic nerve: rhabdomyosarcoma is therefore a diagnostic and therapeutic emergency [16-20], especially since it is a tumor of sudden development [15,16]. Differential diagnoses are numerous and are grouped into two categories: tumoral and non-tumor etiologies [14-21]. Tumor etiologies include cystic tumors (dermoid cysts, embryonal carcinomas), vascular tumors (capillary hemangioma or benign hemangioendothelioma of

infants, lymphangiomas), nerve tumors (orbital neurofibromas, plexiform neurofibroma, optic nerve and chiasm glioma), bone and cartilage tumors (fibrous dysplasia, aneurysmal cysts, juvenile ossifying fibromas, osteosarcomas, chondrosarcomas), histiocytic diseases (Langerhans cell histiocytosis or histiocytosis X, juvenile xanthogranuloma), orbital involvement in blood diseases (lymphomas including Burkitt's lymphoma, tumor localization in leukemia), metastases (neuroblastoma, Ewing's sarcoma), and finally, tumors spread to the orbit including retinoblastoma which still occupies an important place in countries where access to care is more difficult [21,22]. Non-tumoral etiologies are mucoceles, meningoceles, encephaloceles and microphthalmias with cysts, inflammatory pseudotumors, and infections (dacryocystitis, ethmoiditis, orbital cellulitis, orbital abscess).

Conclusion

This observation is atypical due to the orbital location of the tumor, but rhabdomyosarcoma is also a diagnosis that should be considered in order to initiate early treatment for a better vital and functional prognosis.

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