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Orbito-Temporal Hamartoma in a Newborn

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Abstract **Case Report**

We report the case of a male newborn, eleven days old, born in a context of dystocic delivery, but without notion of resuscitation or oxygen therapy. He was referred for protrusion of the left eyeball observed at birth. The right eye examination was normal. In the left eye we found a congenital, axile, irreducible and inflammatory grade III proptosis with corneal dystrophy. The fundus was inaccessible. A computed tomography performed showed a sphenoid heterogeneous expansive process with left temporo-orbital extension with locoregional invasion. The child was referred to the pediatric neurosurgery department where the total tumor extraction was performed and the analysis of the surgical specimen revealed an orbito-temporal hamartoma.

Keywords: Exphthalmia, hamartoma, orbit, newborn.

INTRODUCTION

Hamartomas are benign, congenital, embryonic and multi-tissue tumors [1]. They can occur almost anywhere in the body, including the lungs, heart, skin, brain, breast, or other areas [2]. There have been few reports of the orbit [3]. They occur most often in infants and young children. The age at onset, evolution, local and general examination supplemented by imaging will guide its diagnosis [4]. It is the pathological examination that will specify the histological type [5].

Occurring mainly during embryogenesis, hamartoma is sometimes called dysembryoplasia. Its origin remains poorly understood [6]. Hamartomas can be single or multiple. Symptoms vary depending on location [8] In some areas of the body, hamartoma is asymptomatic, or even regressive over time. In other locations, its development is fraught with complications [7]. Some hamartomas can progress to a malignant (cancerous) form. Multiple hamartomas have a higher risk of malignancy [6]. This complication is rare and represents less than 2% of cases [7]. The treatment is multidisciplinary. Treatment options largely depend on where the tumor is located and whether it is symptomatic or not.

OBSERVATION

We report the case of an eleven-day-old newborn, male, received for protrusion of the left eyeball, observed at birth. The interrogation revealed to us that he was born of a normal pregnancy. The delivery, which took place under medical assistance, was normal, with no notion of resuscitation or oxygen therapy with a normal Apgar score. On ophthalmological examination, the right eye was normal in all these elements. The left eye presented grade III proptosis, axile, irreducible and inflammatory in appearance with filling of the temporal rim. There was a lack of mobility of the globe. Examination of the anterior segment found significant chemosis, diffuse conjunctival hyperemia, corneal dystrophy secondary to exposure keratitis. The fundus was inaccessible. In addition, the general clinical examination carried out by the pediatrician in search of other pathologies was unremarkable. In view of this symptomatology, we evoked the diagnosis of a congenital orbital tumor of the left eye (figure 1). Orbito-cerebral computed tomography showed a sphenoid expansive process with left temporal orbital extension through the orbital fissure. Its density is heterogeneous with predominantly cystic multi-partitioning and with calcifications. It measures approximately 69x29mm. There was also invasion of the optic nerve and infiltration of the oculomotor muscles (Figure 2-4). The management was

carried out in collaboration with the pediatric neurosurgery team and consisted of total extraction of the tumor via the temporal route (figures 5, 6). The anatomopathological examination of the surgical specimen concluded macroscopically with an ulcero-budding tumor with necrotic remodeling of sometimes firm sometimes renitent consistency. Histologically, the fragments presented rabdomyocytes, adipocytes, fibroblasts and chondrocytes without atypia suggesting an orbito-temporal hamartoma. The postoperative course was simple (fig. 6) and regular monitoring is underway to look for complications or tumor relapse. chondrocytes without atypia suggesting an orbito-temporal hamartoma. The postoperative course was simple (fig. 6) and regular monitoring is underway to look for complications or tumor relapse. chondrocytes without atypia suggesting an orbito-temporal hamartoma. The postoperative course was simple (fig. 6) and regular monitoring is underway to look for complications or tumor relapse.

On the ophthalmological level, there is a loss of vision in the left eye by corneal opacity and compressive optic neuropathy and strabismus, the treatment of which has been deferred.

Figure 1: Image of exophthalmos before surgery

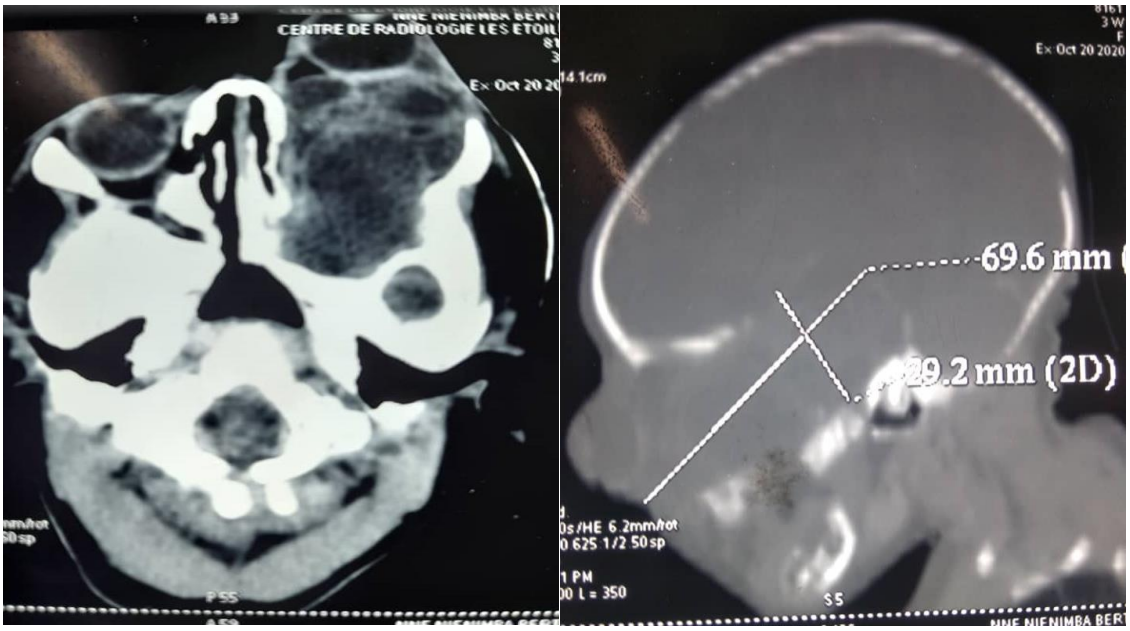

Figure 2: Images of the orbito-cerebral computed tomography before surgery

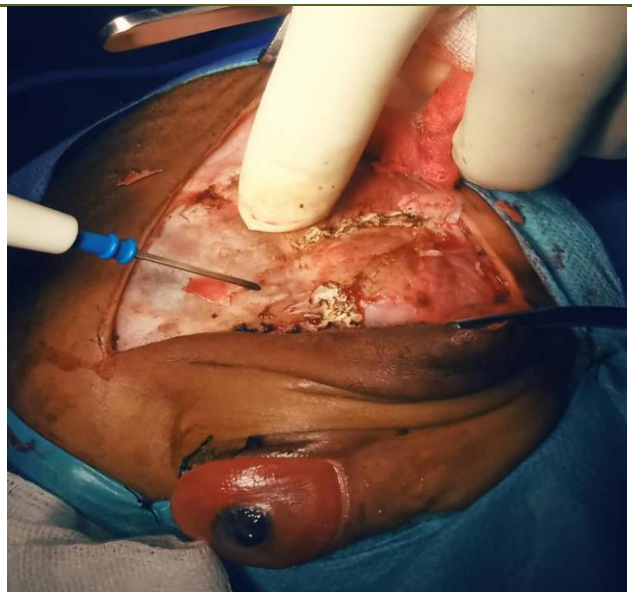

Figure 3: Intraoperative image

Figure 4: Image of the operating room

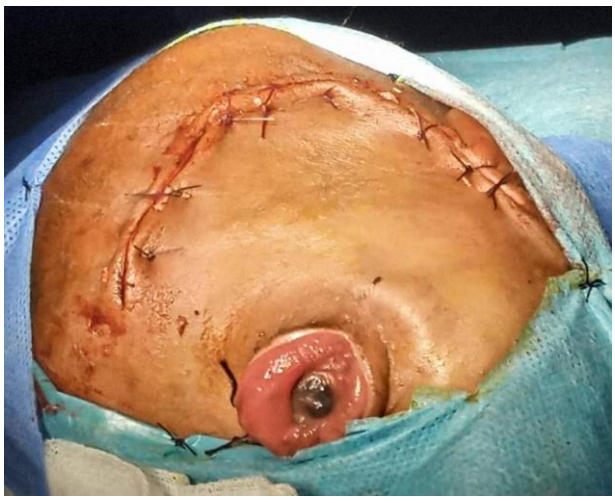

Figure 5: Immediate postoperative image

Figure 6: Late postoperative image

DISCUSSIONS

Hamartomas can develop in many areas of the body.

Since the origin of hamartomas is still poorly understood, no preventive measures have been identified.

They do not all have the same clinical manifestations [6]. In our case the tumor developed at the orbito-temporal level. In the vast majority of cases, proptosis alerts and leads to the diagnosis [10]. However, it should be noted that proptosis in the neonatal period is relatively rare and has various underlying etiologies that can compromise the sight or even the life of the child. Dislocations of the globe following traumaobstetrics,orbital hematoma under neonatal periosteum are responsible for neonatal exophthalmos whose evolution is towards regression. It is accompanied by a lower or upper scleral show, except in tumors of the upper part of the orbit which are associated with proptosis or mechanical ptosis due to associated upper eyelid involvement [1, 11]. It often occurs in infants and young children [4], the case of our patient was observed at birth. It is a non-axile, non-reducible and progressive proptosis. Some hamartomas may remain asymptomatic (with no apparent signs) while others may cause discomfort and/or disrupt function organsdue to the pressure exerted on neighboring organs and tissues [12]. Our patientpresented with tumor-like proptosis, with signs of corneal compression and exposure (chemosis plus keratitis) resulting in total loss of vision in the left eye.

The diagnosis is based on the data of the clinical, radiological and especially histological examination.

There are other locations in the eyeball including the iris of the conjunctiva of the retina.

The management of hamartoma is multidisciplinary and depends on its location, the associated clinical signs, the risk of complications and the patient's condition. In case of symptomsannoyingor risk of malignant evolution, surgery consisting in removing the entire hamartoma is necessary [9]. Our case underwent surgery where the entire tumor was removed by a multidisciplinary team. The pathological examination of the surgical specimen confirmed the diagnosis.

On the other hand, simple medical supervision is put in place if the hamartoma is not associated with any discomfort or complication, thus limiting this risk through early detection, regular medical monitoring and appropriate management [13].

CONCLUSION

Congenital, embryonic tumours; hamartomas are benign tumors, made up of disordered tissue normally present in the organ in which it develops. They can develop in many areas of the body. The orbital location is exceptional. It often occurs in infants and young children; the diagnosis is histological. Resection of the tumor by surgery is most often the only therapeutic alternative. Multidisciplinary collaboration is necessary for its management.

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