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CASE REPORT

Conjunctival myxoma in a pediatric patient: A case report

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ABSTRACT

Conjunctival Myxomais a rare benign neoplasm, with a Mesenchymal origin. We emphasize the importance of excisional biopsies to diagnose indeterminate conjunctival lesions. The patient presents chemosis and translucent conjunctival mass in the right eye, he underwent a next ensive evaluation as well as multidisciplinary management, without evidence of systemic disease associated with conjunctival myxoma was found. The complete excision of the tumor is the optimal treatment as it contributes to the histopathological examination for confirmation. The importance of an adequate diagnosis is to demonstrate the existence of an undetected síndrome, allowing a comprehensive evaluation treatment and multidisciplinary follow-up.

INTRODUCTION

Myxoma is a rare benign tumor of Mesenchymal origin, formed by connective cells that produce abundant amounts of glycosaminoglycans that simulate the umbilical cord matrix (Patrinely, 1983). Previously, it was considered the major primary benign neoplasm in heart frequency, at the moment, it occupies the second place 21.5%, preceded by Rhabdomyoma 50.7%, and can be located in other tissues and organs (Tzani, 2017). In ocular tissues, it can be originated in the orbit, eyelid, conjunctiva, and it is exceptionally present in the cornea, found as localized disease or as a component of the Carney complex, The Mazabraud syndrome and McCune-Albright syndrome (Xiong, 2005). Our patient presents translucent chemosis in the right eye, a histopathological examination was performed and he was diagnosed with Conjuntival Myxoma.

CASE REPORT

The patient is an 11-month-old, with a 25 days evolution palpebral edema in his right eye. The right eye was physically examined and diagnostic shows important chemosis, with in ability to open theeye, with tumor in the tarsalconjunctiva of the right eye of approximately 3x2 cm, tumor with a smooth consistency, smooth edges, well defined, semi-translucent (Figure 1). The patient's face without the presence of lentiginosis, his cardiorespiratory system without compromise. Rhythmic heart sounds have adequate intensity and frequency, lacking the presence of murmurs or acoustic aggregates, dorsal and lumbar region with presence of congenital dermal melanocytosis, (Mongolian spot) of 10 x 10 cm. Diagnostic support and assessment studies were conducted on admission, as well as multidisciplinary management. Myxoma (including cardiac) and endocrine abnormalities were discarded, ruling out the Carney complex. Magnetic Resonance of his orbits and paranasal sinuses were conducted, studies show his right eye ball is smaller than the left eye. The right eye experiences sclera thickening throughout the circumference, a heterogeneous image in the right vitreous appears as a central canal and irregular total retinal detachment are shown. Cysticmass protrudes anteriorly to the right eye. Tomography of the skull is observed changes in the density of soft tissues

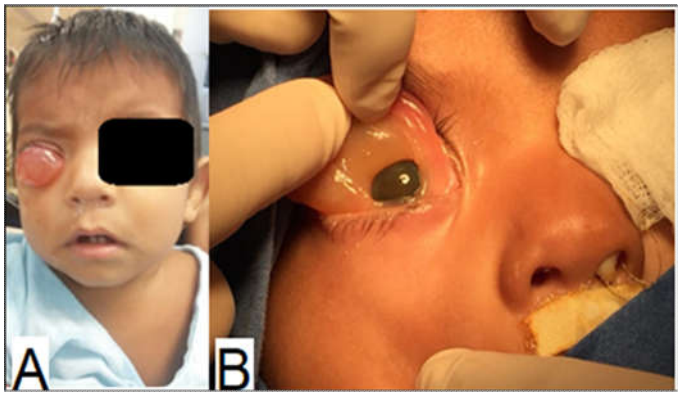

Figure 1. Image shows tumoral lesion Myxoma (A and B). A: Severe Chemosis with exposure of the superior tarsal conjunctiva, and no conjunctiva secretion. Left eye is without alterations. B: Tumor lesion observed during the surgical procedure

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peri ocular right and changes in the density of the vitreous. No proptosis or intraocular calcifications are observed (Figure 2).

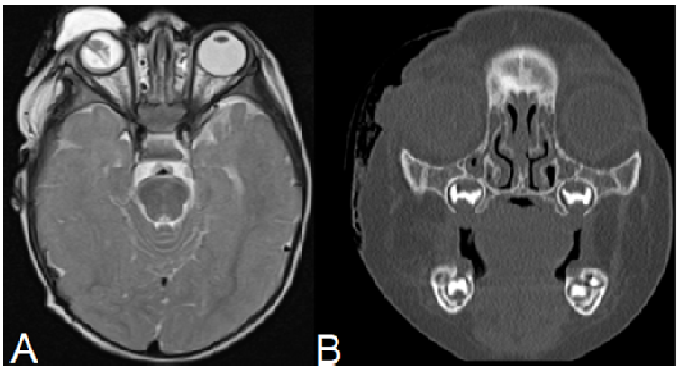

Figure 2. Cranial Magnetic Resonance (A) and Skull Computed Tomography (B). A: In the supraciliary region a discrete hyperintense image is observed in T2, in the Fat Sat it has hypointense behavior that after the administration of contrast has an annular type enhancement of 2mm, which extends to the periorbital and submaxillary soft tissues, as well as the region frontal medial to nasal recess; B: In the tomography of orbit and paranasal sinuses, the evaluated bony structures do not present blastic or lyticlesions

Biopsy of the lesion was performed under sedation and analgesia. In the histopathological examination it was reported: benign conjunctivalneoplasm compatible with myxoma (s100 -, cd34 reactive in vascular wall, myogenin -). Negative for malignancy. (Figure 3). Congo Red specials tain was performed, which was negative.

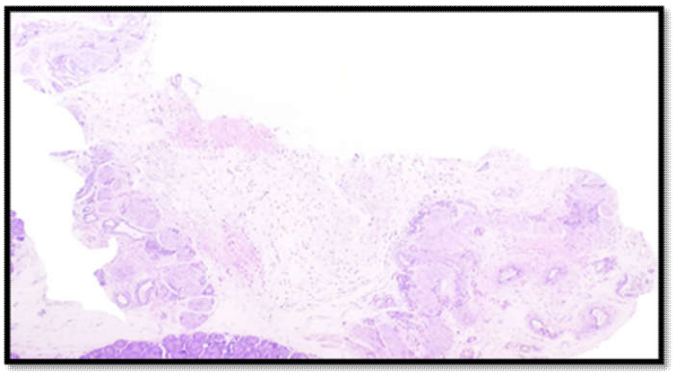

Figure 3. The subepithelial tissue shows a lesión formed by myxoid tissue with medium-sized cells with a starry shape, oval nuclei of fine chromatin, without atypia. It is accompanied by proliferation of arterial and lymphatic vessels, lined by endothelium, of small to médium caliber, with out a typia data. In the interstitium there are some small-sized lymphocytes of mature appearance. The histopathological diagnosis of conjunctival myxoma with cystic change was provided

DISCUSSION

Pure Conjunctival Myxomas are a rare occurrence and only 41 cases have previously been published. This tumor is mainly seen in adulthood, present in an average age of 40 years (range of age 27-51 years), being more frequent in women,(4)with no reported patients in the range age of our patient.

Clinically, Conjunctival Myxomas are a smooth semi translucent or completely translucent, white, pink, pale or fleshy tumors. The tumor can be a solid-cystic, rubbery or gelatinous mass, capsulated or not with acute and well circumscribed edges [3]. Our patient's case presented similar conditions as described above, and the tumor's mass measured 3x2 cm. in diameter. Histologically, Conjunctival Myxomas consists of a tumor matrix that is composed predominantly of Hyaluronic Acid, minor amounts of Chondroitin Sulfates, dispersed vascular structures, and reticulin fibers (3,4). Similar histopathological characteristics were observed in our case. At the moment, the treatment of choice for Conjunctival Myxoma is through surgery with complete excision of the tumor. No tumor recurrence or malignant transformation was observed during the follow-up period of the published cases [5]. The treatment performed in our patient's case was done by completely resectioning the tumor lesion. Given the significant morbidity and mortality of the myxoma associated with the Carney complex and other syndromes, the presence of conjunctival myxoma requires complete systemic as well as multidisciplinary evaluation, the most lethal component of the Carney complex being the development of myxoma and emboli of the heart [6]

Conclusion

The presence of Conjunctival Myxomas is a rare ocular lesion, especially in the pediatric age, the complete excision of the tumor is considered the optimal treatment. The treatment aso contributes to the histopathological examination for confirmation. It is important to give an adequate diagnosis to verify the existence or not of a potentially lethal syndrome, which has not yet been manifested, allowing an exhaustive evaluation, accurate treatment and multidisciplinary monitoring in pediatric patients.

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