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Case Report

Orbito-Palpebral Neurofibroma: A Case Report

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Abstract

Introduction: Neurofibroma is a benign nodular nerve tumor. It may be isolated or associated with neurofibromatosis. Orbital neurofibromas may be associated with lesions of the eyelids, conjunctiva, uvea, sclera and episclera.

Medical Observation: We report the case of a 36-year-old patient who presented with a right superior-internal palpebral swelling that had been evolving for 6 years, with an increase in volume and exophthalmos for one year. The oculo-orbito-palpebral examination revealed preserved right visual acuity of 10/10, a non-axial, painless, irreducible right exophthalmos measuring 23 mm on the Hertel, and a nodular right superior-internal palpebral mass, adherent to the deep plane, painless and with clear contours. An orbito-cerebral CT scan isolated a polylobed right intra-orbital, extra-ocular superior and medial mass measuring 38 mm× 30 mm in axial height, coming into contact with the thickened superior rectus muscle without bony lesions at its contact, in favour of a cystic tumour. An anterior orbitotomy was performed, with excision of the mass in toto. Pathological analysis was in favor of a neurofibroma. Progression was favourable.

Conclusion: Neurofibromas are rare and benign in origin, and surgical removal is for functional and/or cosmetic purposes.

Keywords: *Intraorbital Mass; Exophthalmos; Anterior Orbitotomy; Benign Tumor*

Introduction

Neurofibromas are benign nodular nerve tumors, which may be isolated or associated with neurofibromatosis. Systemic neurofibromatosis exists in 2 forms: neurofibromatosis type 1, caused by mutation of chromosome 17, and neurofibromatosis type 2, caused by mutation of chromosome 22 [1,2]. In its orbital form, it may be associated with lesions of the eyelids, conjunctiva, uvea, sclera and episclera. Orbital neurofibroma is rare, occurring in 1% of orbital biopsies [2]. We report a case of isolated orbital neurofibroma. The aim of this case report is to highlight the clinical, therapeutic aspects of this condition.

Medical Observation

Mr V T, 36, a computer specialist, came to see us because of a progressive increase in a painless mass in the medial part of the right upper eyelid, which had been present for 6 years, with an exacerbation 1 year ago. General and ophthalmological history was unremarkable. Oculo-orbital examination revealed a non-axial, non-reducible, non-painful right exophthalmos with downward and outward displacement of the eyeball, and a non-erythematous swelling located between the medial 1/3 of the right upper eyelid and the medial canthus. Palpation revealed a slightly painful, non-pulsatile mass adherent to the deep plane, with clear contours measuring 10 mm × 15 mm. The

measurement of the right exophthalmos was 23 mm on the Hertel exophthalmometer with limited elevation (Figure 1). On the left, Hertel's ophthalmometry was 17 mm. Uncorrected distance visual acuity was 10/10 in both eyes. The rest of the anterior and posterior segment examination was unremarkable. In summary, the patient presented with a non-inflammatory tumor syndrome of the right palpebra-orbital region, suggesting a benign right orbital tumor. Oculo-orbital CT revealed a right intraorbital, extraocular, polylobed, hypodense mass of sharp, non-vascularized contours measuring 38 ×30 mm (Figure 2). Multidisciplinary management consisted of excision of the right upper-internal palpebro-orbital mass by cutaneous anterior orbitotomy, with anatomopathological examination of the excision specimen (Figure 3). Postoperative medical management consisted of topical and systemic steroidal anti-inflammatory drugs. Pathology revealed a tumoral proliferation of nerve cells grouped in short, undulating bundles with nuclei that were sometimes tapered, rounded or oval, in favor of a neurofibroma (Figure 4). The post-operative course was favorable (Figure 5).

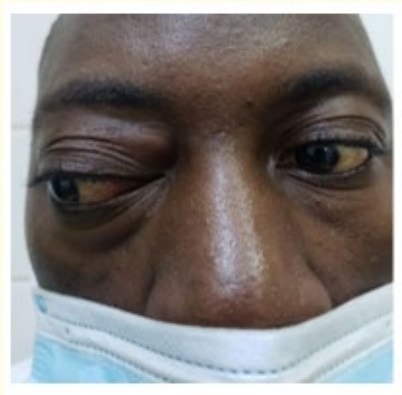

Figure 1: *Right superior. Palpebral mass and exophthalmos.*

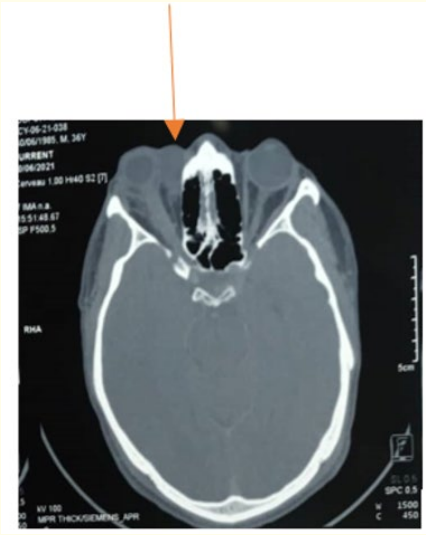

Figure 2: *Extraocular right intraorbital mass polylobed and hypodense.*

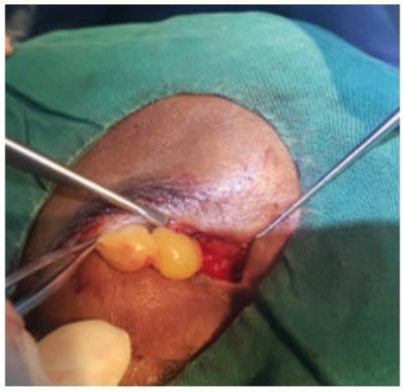

Figure 3: *Removal of a polylobed intraorbital mass.*

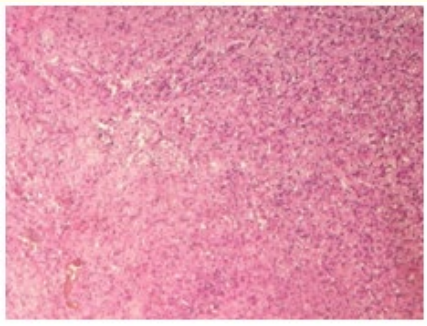

Figure 4: *Anathomopathological results revealing neurofibroma.*

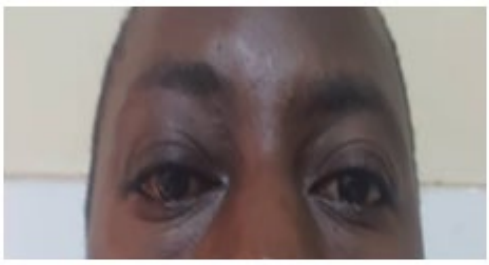

Figure 5: *Month 4 postoperative evolution.*

Discussion

Neurofibroma (NF) results from tumoral hyperplasia of all nerve components, both Schwannian and fibroblastic elements [3].

It is a benign tumor. Orbital neurofibromas may be isolated or associated with systemic neurofibromatosis. In this 36-year-old patient, it was an isolated orbital NF. The localized or isolated neurofibroma is well circumscribed, and typically present in the upper right wall of the orbit, during the 3rd and 5th decades of life with a progressive mass effect Cannon T., *et al*. 2006 [4]. It involves branches of the

frontal nerve [5]. 11 - 28% of patients with localized neurofibromas have a family history or systemic signs of NF1 [6]. In the absence of NF1, malignant degeneration is rare [7]. Clinical diagnosis is made in the presence of an intraorbital mass, which may be associated with exophthalmos, impaired ocular motricity and, rarely, optic neuropathy with reduced visual acuity. The ocular manifestations of NF1, such as Lisch nodules and optic nerve glioma, were not found in our patient. Imaging diagnosis is made using oculo-orbital scanner or magnetic resonance imaging. The diagnosis of certainty is anatomopathological.

Surgical management will depend on the severity of symptoms and clinical signs. The surgical approach is either anterior or lateral orbitotomy, depending on the location and size of the lesion (typically curative) [6]. An anterior orbitotomy with removal of the mass was performed in this patient. The evolution of NF in type 1 NF can have a malignant transformation in 20% and requires clinical monitoring [8]. Isolated orbital NF in the absence of NF1 rarely presents with malignant transformation, but recurrences are possible, hence the need for ophthalmological follow-up.

Conclusion

Orbital neurofibromas are rare benign nerve cell tumours in adults and should also be considered in the presence of an isolated orbital mass. The management of orbital NF is multidisciplinary.

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