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A case of solitary orbital neurofibroma

Simone Ulivieri[1], Davide Luglietto[1], Ludovica Cellini[1], Anna Vaiano[1], Clelia Miracco[2], Antonio Giorgio[3]

[1]Department of Neurosurgery, AOU Senese Le Scotte, Italy
[2]Department of Pathology, Santa Maria alle Scotte Hospital, Siena, Italy
[3]Department of Medicine, Surgery and Neuroscience, University of Siena, Italy

ABSTRACT

Neurofibromas are tumors consisting of a proliferation of peripheral nerves that can affect almost any organ, soft tissue, bone or anatomical site.

We report a case of a 49-old woman with localized orbital neurofibroma, not related to neurofibromatosis. We performed a total resection using Ulivieri's extended lateral approach, with no tumor recurrence at 6-month follow-up.

KEY WORDS: orbital neurofibroma, orbital surgery, Ulivieri's extended lateral approach.

INTRODUCTION

Neurofibromas are tumors consisting of a proliferation of peripheral nerves that can affect almost any organ, soft tissue, bone or anatomical site including, quite commonly, the skin. The systemic manifestation of this disease is called neurofibromatosis type 1 (NF1), which is a widespread and variable disease. Localized neurofibromas of the orbit are relatively uncommon [1]. They account for approximately 4% of orbital tumors and comprise plexiform neurofibromas (2%), localized neurofibromas (1%), and Schwannomas (1%). Schwannomas are generally well defined, often eccentric from the nerve of origin. The main mass of a Schwannoma may contain rare axons which often pass eccentrically through it. Neurofibromas more commonly arise within the nerves and display a more endoneural proliferation, thus expanding and forming an intimate relation to the parent nerve [2].

The symptoms and signs of localized orbital neurofibromas depend on their location in the orbit. Their clinical presentation is similar to the orbital schwannomas and present as a localized orbital mass with slow progressing painless or mildly painful proptosis, globe displacement, impaired extraocular motility, ptosis, sensory dysfunction and, uncommonly, optic neuropathy with decreased visual acuity [3, 4].

We describe here an unusual case of histologically confirmed orbital neurofibroma.

CASE REPORT

A 49-year woman presented with a several-month history of headache and right orbital pain and proptosis. On hospital admission, the neurological examination was normal. Visual acuity, pupillary light reflexes and extrinsic ocular motility were preserved. The fundus examination showed no optic disc edema (Figure 1) and visual evoked potentials (VEP) were normal. A brain MRI with and without

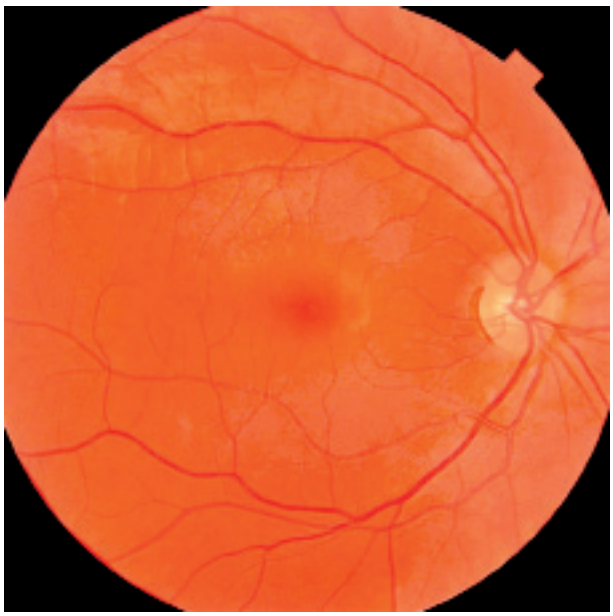

Figure 1. Preoperative fundus examination. No signs of optic disc edema

CORRESPONDING AUTHOR
Dr Simone Ulivieri, Department of Neurosurgery, AOU Senese Le Scotte, Italy, e-mail: simone.ulivieri@tiscali.it

gadolinium showed a well-defined circumscribed intraconal lesion on the right orbit with compression of the optic nerve (Figure 2). The whole pattern prompted us to perform an early intervention. The tumor was surgically removed according to Ulivieri's extended lateral approach [4]. A lateral orbitotomy according to Kroenlein's approach was first performed. At this point, the orbital apex can be reached with a microfresa through the demolition of the lateral wall of the zygomatic bone and the orbital face of the great wing of the sphenoid bone, allowing one to easily treat lesions of the posterior third of the orbit. In this case the lesion was supero-medial, probably originating from the superior branch of the oculomotor nerve. Therefore, after the initial orbitotomy, demolition with a microfresa was cranially directed through the orbital face of the frontal bone. The postoperative course was uneventful without any complication. Postoperative fundus examination was normal and the patient was discharged three days after the operation without any complaint. Histological examination of the lesion confirmed the diagnosis of neurofibroma (Figure 3). A brain MRI examination performed 6 months later revealed no signs of tumor recurrence.

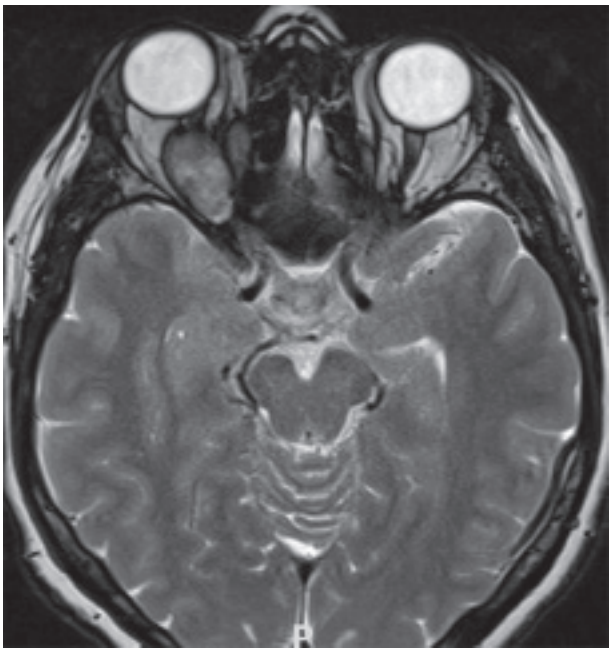

Figure 2. MRI image, in axial orientation, demonstrating the voluminous lesion with defined margins that occupies the right supero-medial intraconal space with dislocation of the optic nerve

DISCUSSION

Neurofibromas are benign, peripheral nerve sheath tumors and, as such, they are composed of variable combinations of Schwann cells, perineural cells, and fibroblasts. Neurofibromas of the orbit are rare and account for 0.6-2.4% of all orbital tumors. There are three types of orbital neurofibromas: plexiform, diffuse and localized. Plexiform neurofibromas,

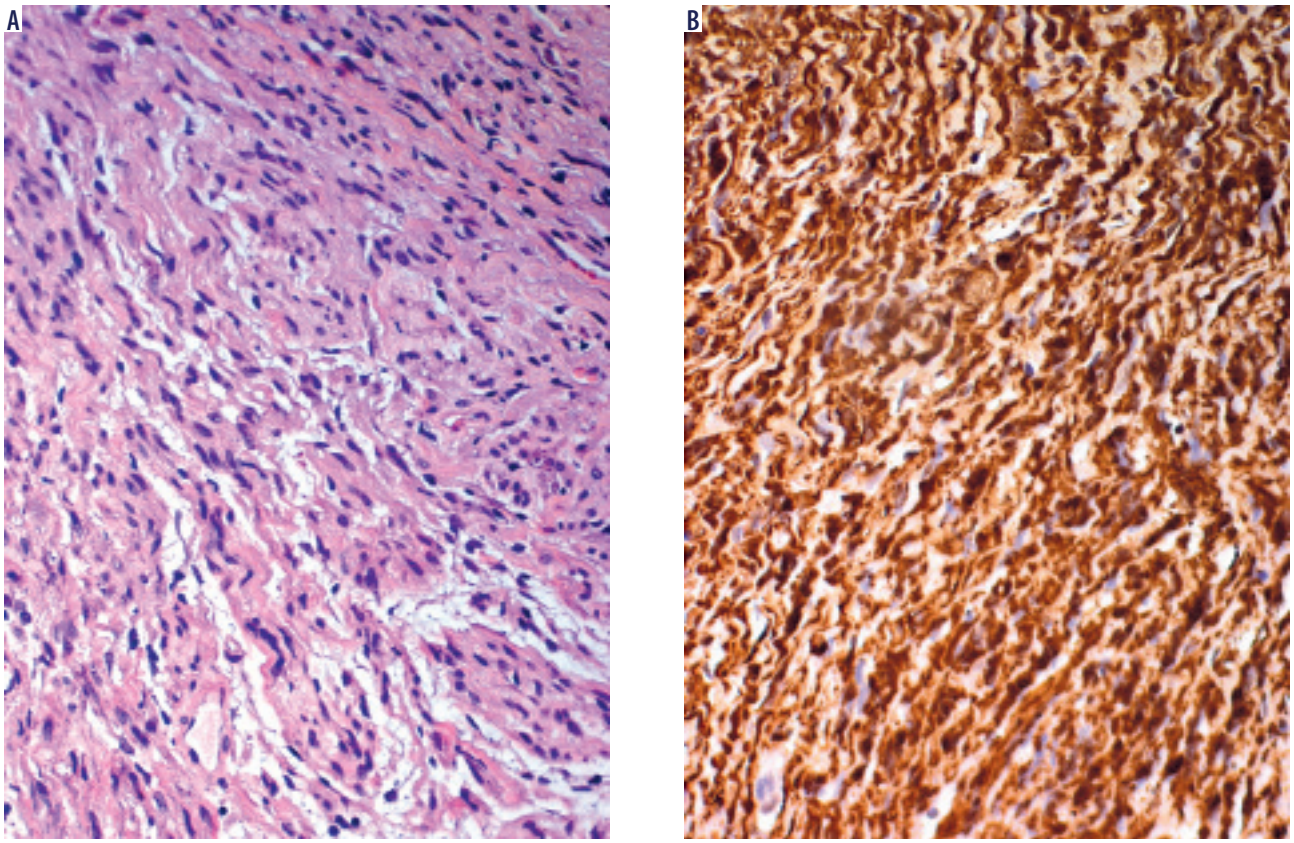

Figure 3. A) Patternless proliferation of spindle cells, with wavy serpentine nuclei, intermixed with wire-like collagen. **B**) Strong, diffuse S-100 immunopositivity. **A**) Hematoxylin and eosin. **B**) Immunohistochemistry, chromogen: diaminobenzidine; original magnification: A, B × 200

the most common orbital subtype, occur exclusively in NF1, become manifest during the first decade of life, and diffusely infiltrate the eyelid and orbital soft tissue. Diffuse neurofibromas are usually the dermal variants; they rarely affect the orbit and are clinically indistinguishable from the plexiform subtype. Both plexiform and diffuse subtypes lack clear margination and tend to be highly vascular. These neoplasms do not respond to medical treatment, and subtotal resection and hemorrhage frequently complicate their surgical management.

Localized neurofibromas of the orbit are uncommon. Although they may arise from either sensory or motor nerves, branches of the frontal nerve are most commonly affected. Presenting typically in the second to fifth decades of life as slowly progressive, orbital soft tissue masses, they induce axial and non-axial globe dystopia. Impairment of visual acuity is typically minimal. Moreover, altered ocular motility secondary to the mass effect may exist.

Localized neurofibromas are relatively well circumscribed and much less vascular, unlike plexiform and diffuse neurofibroma, which are more vascular and diffusely involve the orbital tissues.

The clinical presentation of localized neurofibromas usually depends on the origin and location of the tumor in the orbit. The most common site of localized neurofibromas is the superior orbit, but they may also occur in the inferior orbit. Surgical total resection is the treatment of choice. Surgical approaches and techniques for orbitotomy have been published since its first description in 1841 by Parker [5]. The earliest described techniques of orbitotomy included anterior and lateral transorbital approaches mostly aimed at removing tumors confined within the orbit. The transcranial approach was first introduced by Dandy in 1922 [6]. Endoscopic orbital surgery is also an increasingly useful and less invasive approach. Due to the invasive nature of the transcranial approach, the lateral approach has become the preferred choice [7].

Because of the difficulty of surgical dissection near the apex of the orbit, there is a significant albeit low incidence of surgical complications, including minor leaks of cerebrospinal fluid with subsequent spontaneous cessation; neuroapraxia of motor nerves, possibly due to perioperative edema at the level of the superior orbital fissure; reduction of visual function; fixed mydriasis, possibly due to impairment of blood flow to the optic nerve or ciliary ganglion.

Impaired function of extraocular muscles or elevator palpebrae, often observed in the early post-operative period, generally recovers over several months. In the neurofibromas arising from the sensory nerve the most common complication after surgery is sensory deficit. The lack of a clinically detectable deficit may be due to the fact that these tumors originate from minor branches of sensory nerves, are sited eccentrically with respect to the sensory nerve of origin, or arise from nerves not subserving coetaneous sensitivity such as autonomic nerves, whose involvement may explain some cases with post-operative mydriasis.

DISCLOSURE

The authors declare no conflict of interest.

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