IMAGES, QUESTIONS AND ANSWERS

Right orbital tumor: Which diagnostic orientation? What therapeutic approach to adopt? What definitive diagnosis?

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Case report

A 29-year-old woman, 9 months post-partum, was referred by her ophthalmologist for progressive right visual acuity loss of 4 months’ evolution. Ophthalmological examination found non-reducible right axial exophthalmos with 90\% right- and 100\% left-eye acuity. The right visual field was impaired, with conserved photomotor reflex and eyeball mobility. ENT and somatic examinations were normal. There were no clinical signs of neurofibromatosis.

Orbital MRI found a well-contoured right ovoid mass with a long axis of 23 mm, enhanced on contrast medium injection and with a clear peripheral ring, deforming the posterior pole of the eyeball and compressing the right optic nerve (Fig. 1).
Figure 1  Orbital MRI. A. T1-weighted sequence without contrast injection. B. T1-weighted sequence with contrast injection; arrow points to strong peripheral enhancement. C. T2-weighted sequence without contrast injection.

What is your diagnosis?
Replies

Diagnostic orientation

Large right, apparently benign, orbital tumor, mainly raising the question of preoperative diagnosis.

Treatment approach

The patient underwent modified Krönlein right lateral orbitotomy (Fig. 2). The steps were as follows: C-incision along the outer orbital edge extended along an external canthal fold; exposure of the zygomatic periosteum and orbital rim; zygomatic bone section by circular saw; and exposure of the periorbita. An H-shaped opening of the periorbita allowed exposure of the external rectus and then of the orbital contents.

The operative microscope revealed the tumor and allowed complete monobloc resection and decompression of the optic nerve.

The bone flap was repositioned at end of surgery after suturing the periorbita. Postoperatively, right ocular movement showed impairment, with diplopia, normalizing by 4 months.

 Exploration was conducted with care to conserve the external muscles of the eye and optic nerve, damage to which may result in postoperative mydriasis. Postoperative edema and muscular reaction determined by tumor size account for transient postoperative diplopia or oculomotor disorder. At 12 months follow-up, there was no recurrence, and the esthetic result was satisfactory.

Definitive diagnosis

Definitive diagnosis was anatomopathologic. Macroscopically, there was an ovoid yellowish tumor of 2.4 × 1.8 × 1.7 cm with cystic areas (Fig. 3A). Microscopically, the tumor was encapsulated and constituted by a proliferation of Schwann cells organized in some areas in compact Antoni A bundles and in others in a looser Antoni B configuration (Fig. 3B).

Immunohistochemistry found a 5–7% proliferation index on Ki-67 labeling and positive S-100 protein labeling of all tumor cells (Fig. 3C and D). The diagnosis of benign orbital schwannoma was confirmed. Schwannoma may occur de novo or in association with neurofibromatosis. An orbital location is rare, at 1–6% of tumors as a whole [1]. Orbital schwannoma is often unilateral; the implicated nerve is rarely identifiable. Trigeminate branches (supraorbital and supratrochlear nerves) are the most often affected, although there may also be involvement of the optic, oculomotor, infraorbital, ciliary, lacrimal and zygomaticotemporal nerves [2].

Orbital schwannoma is weakly invasive, with slow evolution that may be accelerated by pregnancy due to iterative intratumoral hemorrhage induced by hypervascularization [3]. MRI is the examination of choice, orienting diagnosis ahead of surgery. Strong peripheral enhancement is pathognomonic [4]. Shen et al. correlated histologic and radiologic findings: Antoni A regions show in intermediate signal on T1 and T2 weighted sequences and are enhanced, whereas Antoni B regions show in hyposignal on T1 and hypersignal on T2, without enhancement [5]. Complete surgical resection is the attitude of choice. Lateral

Figure 2  Right lateral orbitotomy. A. Incision. B. Periorbital exposure. C. Tumor exposure. D. Surgical scar at 2 months.
orbitotomy, as described above, provides a wide operative field for a painstaking search for the implicated nerve and for complete tumor resection. The schwannoma capsule derives from the perineurium of the implicated nerve. Differential diagnosis concerns cavernous hemangioma or isolated neurofibroma. With appropriate treatment, prognosis is very good. Recurrence is rare, and mainly due to incomplete resection, schwannomatosis or plexiform schwannoma.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References