Intra Orbital Schwannoma

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Introduction

Solitary intra orbital schwannomas are rare tumors. The diagnosis is made by an MRI and can be mistaken for cavernous angiomas. The treatment for solitary intra orbital schwannomas is exclusively surgical. The prognosis is generally good. We present the first case of intra orbital schwannoma operated in our department.

Observation

We report a case of a 54 year old patient ZN, with past medical history of Diabetic Mellitus and underwent a thyroidectomy. She presented with left progressive, axial, painless exophthalmos, without signs of inflammation for 10 years, without affecting visual acuity. Clinical examination revealed patient with a grade III exophthalmos, visual acuity 10/10 in both eyes, normal visual field, normal fundoscopy examination and normal ocular muscle movements.

Orbito-cerebral CT scan showed a left intra orbital fusiform tissue formation behind the ocular globe compressing the optic nerve, heterogeneously enhanced by contrast, measuring 25x22x19 mm.

MRI showed a left well circumscribed, intra conical intra orbital mass with regular margins, appearing hypo-intense in T1 sequence and heterogeneous in T2 sequence, with a hyper-intense collarette. This mass displaced

1. Above: the ocular globe producing a grade III exophthalmos.
2. The optic nerve above and interiorly without signs of infiltration.
3. Laterally: oculomotor muscles.
4. Exteriorly: lacrimal gland, moderate homogeneous increase in volume thus protruding, where the aspect evoked a cavernous angioma.

The patient was operated by a fronto-orbital approach with good post-operative outcomes. Histopathological results revealed a benign intra-orbital schwannomas.

Discussion

Orbital schwannomas are rare orbital tumors. They represent 2-5% of intra orbital tumors. They are generally painless slow growing tumors in a period of years. It is an extra optic tumor and the visual acuity is usually conserved except in the case of compression of the optic nerve by significant tumor volume and if it is posteriorly located; it can be associated with Von Recklinghausen disease.

Intra orbital schwannomas generally arise from intra orbital sensory nerves. The most frequent being the supra orbital and supratrochlear branches of the frontal nerve [1] evoking also a hypoesthesia in their territory, which differentiates them from cavernous angiomas. Other nerves that can be the origin of intra orbital schwannomas are notably the infra orbital nerves [2] or the naso ciliary nerve [3].

Intra orbital CT scan shows an isodense, well circumscribed, homogeneous lesion enhanced by contrast. MRI T1 sequence shows a well circumscribed lesion in the orbital fat and iso-intense with the muscle. A fine peripheric annular contrast enhancement seen in 29% of cases is pathognomonic of the diagnosis [4]. In T2 sequence the tumor appears hyper intense or in alternating Hypo and Hyper signal.

Cavernous hemangioma is the only differential diagnosis, but on the MRI we see a well circumscribed homogeneous signal. Contrast enhancement of hemangiomas starts at a certain point and then extends in an oil drop manner in a homogeneous fashion all over the tumor; contrary to schwannomas where the contrast enhancement is taken at once throughout the tumor [5].

Anatomopathologically, it is a solid encapsulated tumor and is composed of fusiform schwannoma forming the palisades and compact bundles (Antoni A) or in a rarer configuration without a particular arrangement (Antoni B). Cystic forms are rare [6-9]. The cystic transformation of the tumor is the consequence of intra tumoral necrosis and intra tumoral hemorrhagic phenomena [8]. The only effective treatment is surgical which gives a definitive cure without recurrence of total excision is achieved. The surgical
approach depends essentially on tumor location and volume, in our case we used a supero lateral orbitotomy.

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