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Sir,
Lipomatosis of the Orbits: possibly a form of Madelung's disease

Madelung's disease is a systemic disorder, which is typically characterized by symmetrical deposits of adipose tissue in the neck and upper back. In 1888, Madelung collected a series of cases outlining the macroscopic structure and clinical features of this disease. It is more common in males and associated with high alcohol consumption.¹ In this report, we present the clinical and radiological manifestations of Madelung's disease in the orbits, which to our knowledge has not been described before.

Case report

A 49-year-old Russian male was referred to the orbital clinic with prominent eyes. He had no previous ocular history. Three months before this he was diagnosed with Madelung's disease when he presented with neck swelling secondary to fatty infiltration. He underwent bilateral neck dissections with histology revealing well-differentiated adipose tissue. He denied excessive alcohol consumption although his gamma-glutamyl transpeptidase was raised at 202 (normal range 2–50).

On examination, his visual acuity was 6/6 with normal colour vision on Ishihara test plates for both eyes. Hertel exophthalmometry revealed proptosis of 23 mm bilaterally and marked periorbital soft-tissue fullness (Figure 1). He had full range of eye movements and his remaining eye examination was normal. His thyroid function tests were normal. A computerized tomography

Figure 1 (a) Photograph of patient's eyes in the primary position, demonstrating bilateral proptosis and periorbital fullness secondary to adipose deposition. (b) Photograph of patient's neck with pathological fat deposition.

Figure 2 Axial computer tomography scan of the brain and orbits, demonstrating symmetrical straightening of the optic nerves secondary to orbital fat deposition and proptosis.

scan of the orbits demonstrated symmetrical orbital fat deposition with straightening of the optic nerves (Figure 2). In view of the worsening nature of the disease and straightening of the optic nerves indicative of risk of compromise, he underwent bilateral medial orbital wall decompression and fat excision. Post operatively, he achieved a 2 mm reduction in proptosis bilaterally. Histology of excised fat demonstrated mature adipose tissue consistent with Madelung's disease.

Comment

Madelung's disease is characterized by multiple non-encapsulated lipomas, which can enlarge significantly, resulting in dyspnoea and dysphagia.² The etiology of the disease remains unknown, but an abnormal lipogenesis induced by catecholamines has been observed and is often associated with liver dysfunction due to alcoholism.³

Bilateral late-onset lipomatosis of the orbits also occurs in Graves' disease and has also been reported following prolonged administration of oral corticosteroids.⁴ Interestingly, the phenotype of orbital fibroblasts is distinct from pre-adipocyte fibroblasts elsewhere, and it is this characteristic that is thought to contribute to the selective involvement of the orbit in Graves' disease; yet, in Madelung's disease the orbit also appears to be a feature.⁵ Treatment is surgical removal of excess adipose tissue where it is affecting the aerodigestive tract, causing psychological distress and may now be extended to include compromising optic nerve function.

Conflict of interest

The authors declare no conflict of interest.

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Sir,

Recurrent giant orbital apocrine hidrocystoma

We report a case of recurrent giant hidrocystoma that demonstrates the importance of intact excision of these lesions.

Case report

A healthy 46-year-old woman with a few months history of painless swelling of the right lower lid. There was no history of impaired vision, diplopia, watering, discharge, or prior trauma. Ophthalmological examination revealed a soft, non-reducible, medial lower-lid swelling with normal overlying skin. The eye was displaced superiorly and laterally. Valsalva test was negative and ocular motility was full. An orbital CT scan showed a large, cystic mass of about 18-mm diameter occupying the infero-nasal orbital quadrant. Surgical excision via conjunctival approach revealed a very thin-walled cystic mass that ruptured during the procedure with the release of clear fluid. The histopathological examination of remnants of cyst wall was inconclusive.

The patient represented 2 years later with a recurrence of the cyst, width 17.5-mm diameter on MRI scan (Figure 1), abutting the globe in the right inferior medial orbit similarly to that seen previously. The cyst was excised intact. Histopathological analysis revealed a benign apocrine hidrocystoma. No further recurrence of the lesion was seen on repeat MRI at 6-months follow-up.

Comment

Hidrocystomas are benign sweat-gland tumours that are usually small and are typically found in the head and neck region, especially the eyelid, but other locations including the chest, axilla, palms, and penis have been described.¹ Histologically, two types of hidrocystoma, eccrine, and apocrine are recognised reflecting the cells of origin. Spontaneous orbital apocrine hidrocystoma has been reported only once,² a 10-mm cyst located beneath the orbital roof. Giant hidrocystomas are very unusual, the largest reported by Laktaoui *et al.*³ being a 2-cm eccrine cyst on the eyelid.

The pathophysiology of hidrocystomas is uncertain. It has been hypothesised that sequestration of epithelial cells at the embryonic stage could lead to the condition.⁴ Bone remodelling or orbital enlargement is often present in long-standing lesions of the orbit but these features were absent in our patient. Traumatic implantation of epithelial cells into deeper tissues, including the orbit,⁵ has also been described but a history of trauma was absent in our patient.

Figure 1 Orbital MRI Scan. Arrow shows a giant hidrocystoma (17.5-mm diameter) abutting the eyeball medially in the orbit.