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Rare multiple orbital localizations of sarcoidosis

A case report

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Summary. We report a case of bilateral orbital sarcoidosis without other systemic lesions. Steroid therapy did not improve the clinical status of the patient.

Key words: Computed tomography – sarcoidosis – orbital masses

Cranio-orbito-facial sarcoidosis is well known. The granulomatous mass lesions involving the calvarial [36, 37] meninges, cranial nerves, cerebral hemispheres, cerebellar and mesencephalic structures [2, 8, 10, 13, 17, 20, 23, 27, 29, 31, 33], and pineal gland [32] especially have been reported in the literature. The orbits and their content are occasionally affected, especially the optic nerve or its sheath [5, 11, 15, 16, 18, 19, 26, 34], and the ocular bulb, that is, more commonly, the retina [7, 12, 21].

Conjunctival sarcoid granuloma is very common and conjunctival biopsy with histologic examination of the grasped fold was therefore suggested.

We report a case showing several intraorbital bilateral sarcoid masses not in the usual places and without extraorbital localizations.

Case report

A 60-year-old man, showed severe proptosis, worse on the right, bilateral blurred vision and subconjunctival granulomatosis, which increased over nine months. The past medical history included bronchopneumonia and chronical rhinopharingytis.

Visual acuity was limited to 6/10 in the right eye and 8/10 in the left. The visual fields were normal. An afferent pupillary defect was present in the right eye. The fundus showed a bilateral peripheral retinal dystrophy and moderate swelling of the right optic disk.

The neurologic and psychiatric examination was normal.

Routine laboratory tests were normal. The erythrocyte sedimentation rate was 44 mm/h. A chest roentgenogram disclosed tubercular calcification in the right mid-lung field without hilar adenopathy. Skull films were normal; in particular there were neither hyperostosis nor lytic lesions of the anterior clinoids.

An orbital CT showed (Fig. 1) bilateral proptosis, of about 1 cm in the right eye and 5 mm in the left, caused by three retrobulbar masses. All the lesions showed increased density and irregular outline. The more lateral lesion in the right eye covered the supero-latero-posterior part of the ocular bulb between the optic nerve and the lateral rectus muscle; this lesion infiltrated into the lacrimal gland and the superior and lateral recti muscles. The second lesion infiltrated into the medial rectus muscle and grew upwards for about 1 cm.

The pathologic mass, in the left orbit, smaller than in the right, covered the supero-postero-medial part of the ocular bulb between the optic nerve and the medial rectus muscle. All the lesions disclosed a marked contrast-enhancement, increasing by 10 HU.

The diagnosis was made difficult by the absence of other clinical or radiological systemic findings. However, the conjunctival biopsy showed noncaseating epithelioid granulomas, compatible with sarcoidosis, thus resolving this difficulty.

The patient was given steroid treatment without subsequent improvement.

A second CT examination was made 16 months later (Fig. 2). It disclosed, in the right orbit, a marked increase of the medial mass which infiltrated the me-

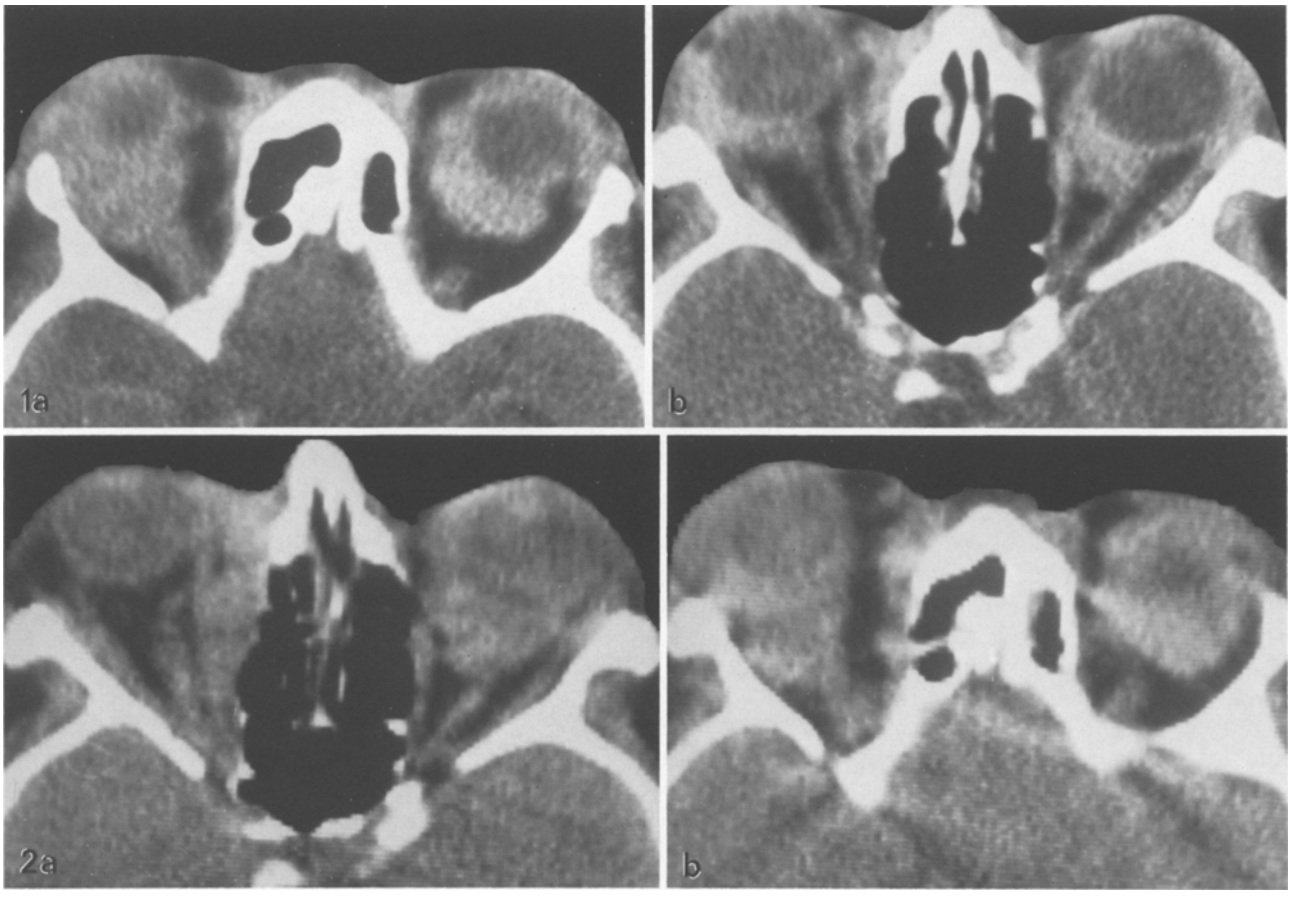

Fig. 1 a and b. After contrast enhancement. The lateral mass in the right orbit infiltrates the lacrimal gland. The mass in the left orbit grows between the medial rectus muscle and the optic nerve (**a**). The medial mass in the right orbit infiltrates the medial rectus muscle (**b**)

Fig. 2 a and b. The medial mass fills the medial part of the right orbit and causes a marked increase of the proptosis, dislocating the retro-orbital fat (**a**). The other lesions has slightly increased (**b**)

dial rectus muscle and filled the medial part of the orbit.

The pathologic mass in the left orbit was slightly increased.

Discussion

The case discussed here is different from those reported in the literature for the following reasons: (a) The orbital localizations remain isolated without other systemic lesions; (b) the presence in the orbits of three large sarcoid masses which caused bilateral proptosis and contrast with the absence of systemic disease; (c) the unusual localizations away from the optic nerve which is commonly affected (in which case the differential diagnosis between neoplastic lesions of the optic nerve and sarcoid masses is very difficult) [9, 28, 30, 35]; (d) the granulomatous masses included, in the right orbit, the lacrimal gland and the medial rectus muscle; in the left eye, a single mass which had grown between the optic nerve and the lateral rectus muscle; (e) the CT images were similar in all the localizations and not completely in accordance with those found by other authors [3, 12], i.e., the tissue masses had increased density with marked contrast enhancement and irregular outline; (f) there was neither hyperostosis nor osteolysis of the orbital bones notwithstanding the peripheral position of some lesions [1, 24, 25]; (g) steroid treatment did not improve the clinical status contrary to the experience of other authors [6, 14].

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