ORBITAL VARIX (Case Report)

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SUMMARY :

A patient with an orbital varix manifesting with proptosis, ophthalmoplegia and partial ptosis is presented. The lesion was unsuspected during computerized topographic examination and verified as a venous varix on pathological examination. Detail clinical manifestation and treatment are discussed.

KEY WORDS :

Orbital varix, ophthalmoplegia, Proptosis

Orbital varices are uncommon lesions that characteristically produce unilateral positional exophthalmos and proptosis1-7. The condition is characterized by pronounced and rapid protrusion of one eye when venous stasis is induced by bending the head forward, cowering or forcibly turning it, hyperextension of the neck, couqhing, forced expiration or pressure on the jugular veins 2,7,10. There has been considerable speculation on the aetiology. Recent advances in neuroimaging techniques now make it possible to diagnose orbital lesions precisely and select the best treatment procedures for this disease 3,8-10.

We present a child with a right orbital varix and phlebolith who presented with ophthalmoplegia and proptosis.

CASE REPORT

This-8-year old girl was admitted to hospital complaining of proptosis and double vision in the right eye. She first noticed double vision two months ago. Neurological examination revealed absence of right eye movements with lateral gaze and partial oculomotor palsy with proptosis on the right eye. There was no conjunctival congestion or eyelid swelling related to head position or valsalva manoeuvre. Auscultation of the periorbital region was negative. Ocular tension and fundoscopic examination were normal. Orbital and cranial computed tomography(CT) scan disclosed a moderately high density mass occupying the right orbital cavity, remarkable enlargement of the mass through the superior orbital fissure and involvement of the cavernous sinus (Fig.1). Right superiolateral orbitotomy was performed in July, 1990. After incision of the periorbita the superior levator palpebral muscle was retracted laterally then the periorbital fat was removed and a major portion of the varix, which localized between the right side of the optic nerve and the lateral rectus muscle, was exposed using a microdissection technique. After coagulation the varix was completely removed. CT obtained a mounth after the operation showed complete disappearance of the varix (Fig.2). The right oculomotor and abducens palsies completely subsided and the patient returned to school. She had no functional deficits when last examined in August 1990.



Fig. 1 : Preoperatif CT apperance.



Fig. 2 : Postoperatif CT after total removal of the orbital varix.

DISCUSSION

The typical clinical symptom of an orbital varix is the dramatic appearance of intermittent proptosis which depends upon the patient's postural changes. The proptosis may soon disappear when venous congestion is relieved and commonly results in enophthalmos when head is erect 1-3,8,10. Sudden episodic attacks of severe proptosis sometimes accompained by pain and diplopia lasting from a few seconds to several days occure. These ocular and orbital symptoms, indicating a possible varix, are relatively unusual 6.7. In some patients proptosis gradually prognesses over the years. Spontaneous improvement is rare. Ophthalmoplegia may also result from repeated severe attacks 3,6,7.

In the presented case the clinical manifestation of the orbital varix was unusual. The oculomotor and abducens palsies, exophthalmos and ptosis developed gradually over several weeks. On no occasion our patient's clinical picture was changed by postural changes. It is reasonable to assume that the neurological symptoms were due to a thrombosis formed by longstanding blood collection. The sudden appearance of abducens palsy then gradually progressive proptosis accompanied by partial oculomotor palsy in the right eye were probably due to thrombosis.

In the literature sudden orbital and ocular symptoms suggested that the thrombosis might ha-

ve attacked the vascular wall, causing first lameller fibrosis then central necrosis and finally resulting in the formation of a stone that was isolated from the vascular wall 6.7. The phlebolith lodged at the neck of the varix presumably occluding the venous channel, eventually enlarged the varix and compressed the surrounding intraorbital structures 1.6.7.10. Ophthalmoplegia probably occured as a result of the nerves being compressed by the engorged varix 6.7.

The same mechanism may explain the clinical features of our case.

According to Lloyd7 enlarged intraorbital venous channels are classified as primary and secondary. Primary intraorbital varices are in most instances true congenital venous malformations. Traumatic varices and varices associated whith orbital haemanjioma are also considered primary. Whereas enlargement of the intraorbital veins caused by shunt in cases of carotid cavernous fistula or AVM are considered secondary7. In our case the orbital varix is considered primary.

Differential diagnosis between tumours and true vascular abnormalities may be nearly impossible on clinical grounds alone, orbital venography is essential for an accurate preoperative diagnosis. Also CT image finding added to the typical clinical picture makes the diagnosis of orbital varix highly probable.

In our case the clinical manifestations were unusual for orbital varix. The initial axial and coronal CT scan demonstrated an irregular orbital apex mass which enhanced homogeneously after intravenous contrast administration suggesting a tumoural mass. Treatment depends on the size of the varix, the duration of the proptosis and the degree of disease progression. Surgical intervention should be recommended when repeated episodes of exophthalmos threaten visual function or cause intractable pain, surgery may also be required when trombosis or heamorrhage suddenly occur 1,3,4,6,10,11.

In our case acute trombosis presumably took place and caused the ocular symptoms.

Orbital varix may be associated with venous malformations elsewhere in the body. Vascular abnormalities may be present in the conjunctiva or eyelids and localised venous dilatations on the forehead and scalp are also reported 2,4,11. In our case there was no other vascular abnormality elsewhere in the body.

Injection of sclerosing agents into the orbit, aspiration of the retrobulbar space, ligation of the superior ophthalmic vein, artificially-induced electrical current and total removal of the lesion are the surgical modalities of orbital varix 1,3,6,11. Correspondence : Doç. Dr. Kaya AKSOY M.D. Uludağ Üniversitesi Tıp Fakültesi Nöroşirürji A.B.D. 16069/BURSA Telephone: 9-(24)-605050-5/99

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