ORBITAL LYMPHANGIOMA: CLINICAL FEATURES AND MANAGEMENT

KALISA P.,**, VAN ZIELEGHEM B.***, ROUX P.**, MEIRE F.**

ABSTRACT

Purpose: To report 5 cases of orbital lymphangioma**.**

Methods: All patients underwent ophthalmological, clinical and neuroradiological evaluation.

Results:

At presentation all patients (4 children and one adult patient) had unilateral proptosis. Restricted eye movement was present in 3 patients, ptosis and amblyopia in 2. Compressive optic neuropathy with visual loss was noticed in one patient. Palatal localization was found in 2 patients, maxillofacial localization in one. Associated intracranial vascular anomaly was present in one patient.

Three patients who underwent surgery, developed recurrences.

No regression of lymphangioma was noticed nor with Interferon, nor with steroids.

Conclusion:

Lymphangioma has to be included in the differential diagnosis of childhood proptosis.

Extraorbital localization, under which associated intracranial developmental venous vascular anomaly, has to be searched for.

In most of the cases conservative treatment is mandatory.

RÉSUMÉ

But: Rapporter 5 cas de lymphangiome orbitaire.

Méthode: Tous les patients ont subi une évaluation ophtalmologique, clinique et neuro-radiologique.

Résultats:

Tous les patients ont présenté une exophtalmie unilatérale.

Une limitation de la motilité oculaire était présente dans 3 cas, un ptosis dans 2 cas.

Une amblyopie de l'œil atteint fut observée dans 2 cas.

Un cas a présenté une neuropathie optique compressive. Une localisation extra-orbitaire (palatale dans 2 cas, maxillofaciale dans 1 cas) et une anomalie vasculaire intracranienne (dans 1 cas) ont été rétrouvées.

Les 3 cas ayant subi un traitement chirurgical ont tous récidivé.

Aucune régression n'a été observée dans les cas traités à l'Interferon ou à la cortisone.

Conclusion:

Le lymphangiome orbitaire doit être inclus dans le diagnostic différentiel de l'exophtalmie des jeunes enfants.

Les localisations extra-orbitaires et notamment la présence d'une anomalie vasculaire intracranienne doivent être recherchées.

Dans la plupart des cas un traitement conservateur est recommandé.

KEY-WORDS

proptosis, orbital lymphangioma, orbital mass, intracranial venous vascular anomaly.

MOTS-CLÉS

exophtalmie, lymphangiome orbitaire, masse orbitaire, anomalie vasculaire intracranienne.

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INTRODUCTION:

Lymphangioma is a benign vascular tumour which is probably congenital, slowly growing and may not become clinically apparent for months and for years. The tumour may affect the conjunctiva, the lids and the orbit. Associated similar extraorbital lesions include facial and palatal cystic lesions. Intracranial vascular anomalies have been reported (3, 4, 11). We report 5 cases from which the clinical features and management are discussed.

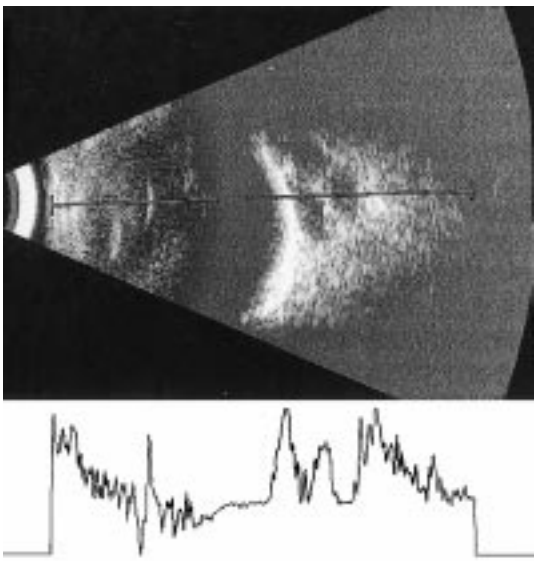

Fig 1A. (-scan echography: displays low reflective, multicystic lesion within the orbit.

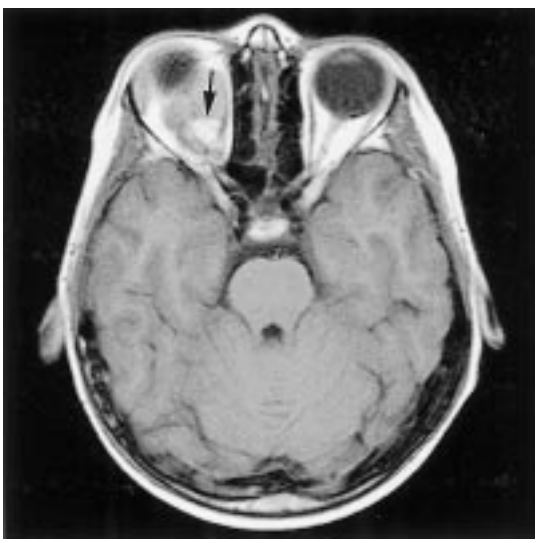

Fig 1B. MRI-T_1 axial: multilobulated intra-orbital mass with internal hyperintense signal representing a recent bleeding.

Case reports

Case 1

In September 1999, a 5 year old boy presented with a sudden onset of right proptosis (4 mm

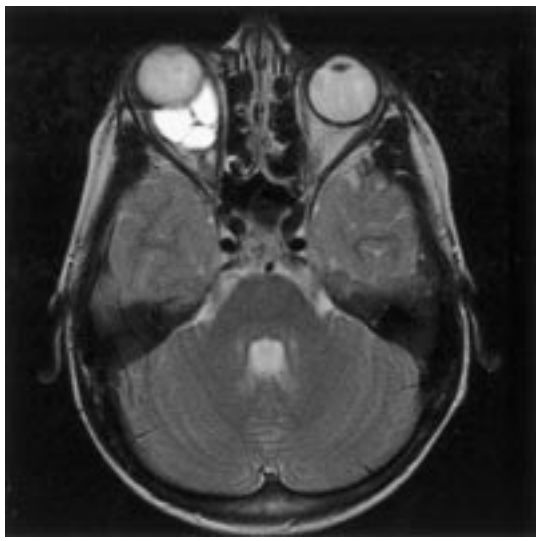

Fig 1C. MRI-T_2 axial: the septations within the tumour are better visualized.-

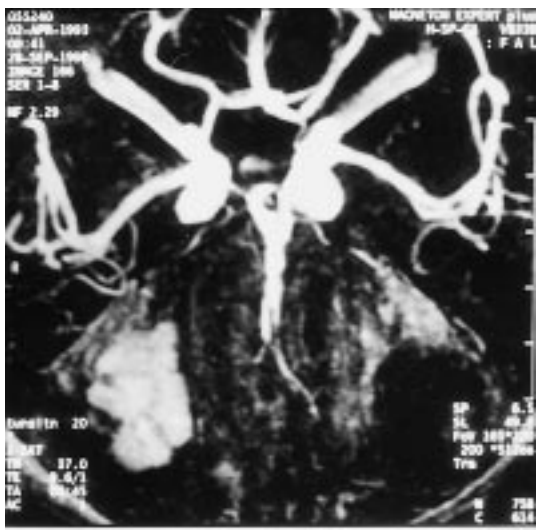

Fig 1D. MRI angio shows no abnormal feeder vessels toward the tumour.

on Hertel) associated with mild limitation of the motility in upward gaze. There was no diplopia. The visual acuity was 10/10 in both eyes. Biomicroscopy and fundoscopy were normal.

Echography documented a lobulated retrobulbar mass with low internal reflectivity (Fig. 1A).

MRI T_1 and T_2 weighted images showed a hyperintense multilobulated intraconal and extraconal cystic lesion, extending between the orbital roof and the superior rectus muscle (Figs 1B and 1C). On MRI angio the tumour showed no connections with the orbital veins and was differentiated from orbital varices (Fig. 1D).

During the 2 year follow-up period 2 recurrent episodes of a sudden increase of the proptosis was observed. The first recurrence occurred shortly after the boy suffered of an upper respiratory tract infection.

The second episode of proptosis was more important (8 mm on Hertel) and related to an haemorrhage within the tumour with subconjunctival bleeding and diplopia. The visual acuity remained 10/10 and it was decided to manage conservatively.

Case 2

In 1997, a 6 year old boy presented with 5 mm proptosis of the right eye and moderate ptosis of the right eyelid (Fig. 2A). Visual acuity was 6/15 in the affected eye. Limited elevation of the right eye without diplopia was present. On biomicroscopy, a gelly-like swelling of the conjunctiva involving the superior nasal quadrant was noted. On oral inspection palatal cystic lesions were observed (Fig. 2B).

Computer tomography showed an intra- and extraconal lesion with lateral displacement of the medial rectus. The diagnosis of lymphangioma was made on clinical ground and a 6 month follow-up was adviced.

In November 1998, he was admitted in the department of paediatric surgery for a right cheek swelling, for which a resection biopsy was performed. Pathological examination was considered normal.

In June 2000, a second acute onset of right proptosis was noticed. This case was managed conservatively and the proptosis remained equally.

Fig 2A. Notice a right proptosis due to an orbital lymphangioma.

Fig 2B. Palatal blood-filled cystic lesions.

Case 3

In 1985, an 18 year old female presented with a history of left proptosis. At the age of 14 a retrobulbar extraconal mass was documented on CT-scan and on MRI (Figs 3A, B and C). The tumour was resected by a transfrontal aproach.

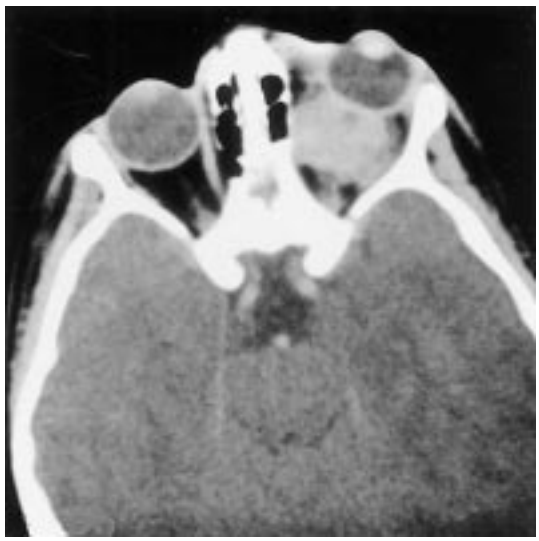

Fig 3A. CT-scan: left-sited intraorbital lymphangioma.

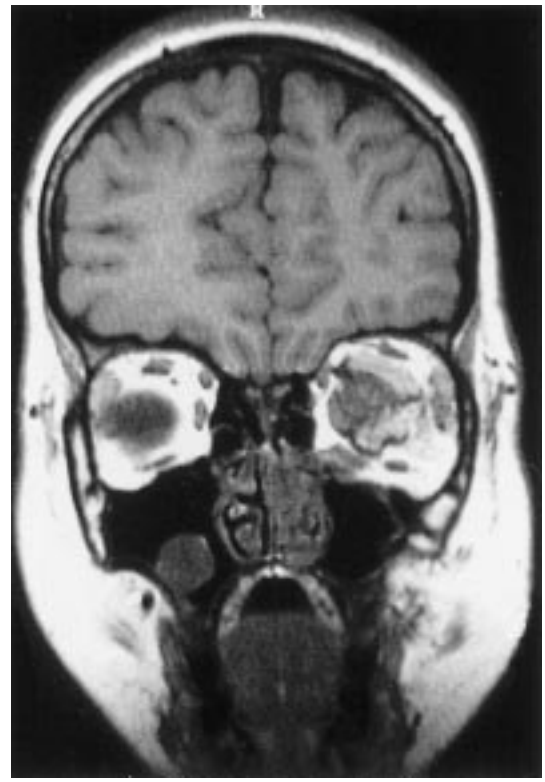

Fig 3B. MRI-T_1 coronal: intraorbital lymphangioma.

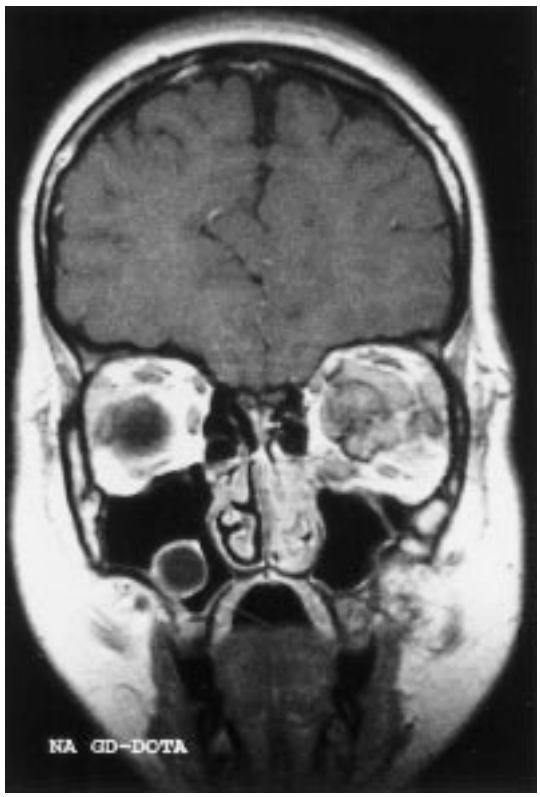

Fig 3C. MRI-T_1 after gadolinium: shows the absence of contrast enhancement

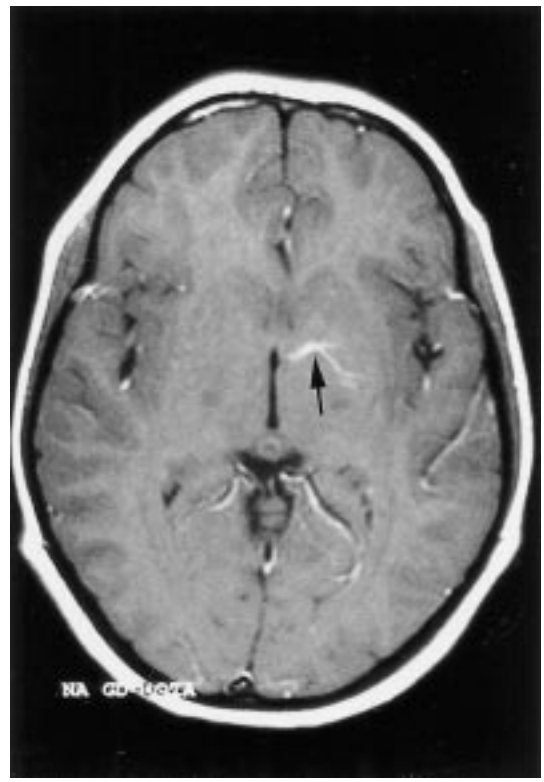

Fig 3E. Intracranial developmental vascular anomaly.

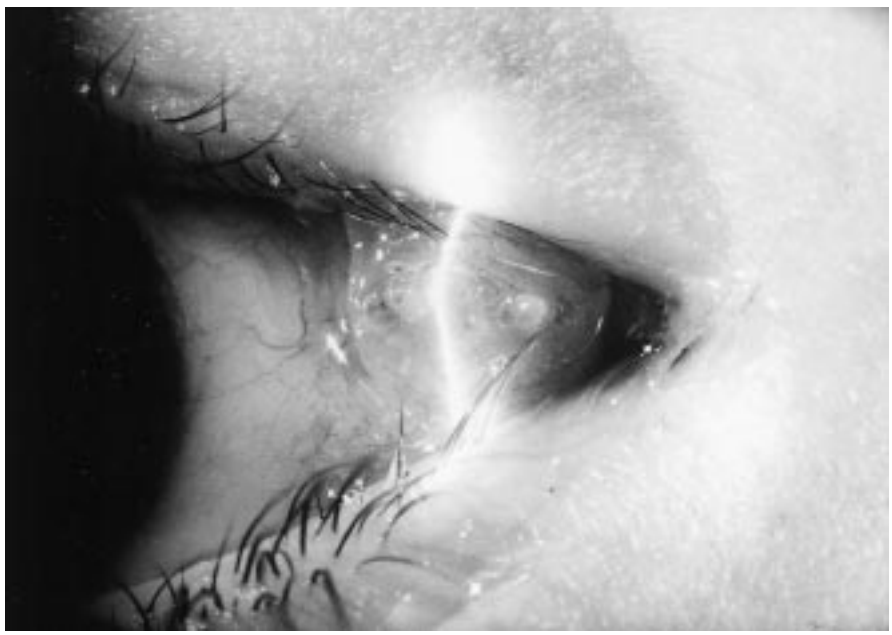

Fig 3D. Conjunctival multicystic lesion, characteristic for lymphangioma.

Pathological diagnosis was consistent with lymphangioma. Postoperative complication included a third nerve palsy and decreased visual acuity (4/10) in the affected eye.
She subsequently underwent resection of a palatal lymphangioma (March 1991) and a conjunctival tumour (February 1995) (Fig. 3D). Despite resection, recurrence of conjunctival and palatal multicystic lesion was observed after a few months. CT-findings suggested that there still remained an orbital component of the tumour.
In December 1995, a multidisciplinary team proposed to treat the patient with Interferon therapy but no improvement was observed. In August 1997 an intracranial developmental vascular anomaly (Fig. 3E) was disclosed and neurosurgical resection of the venous angioma, associated with cavernous angioma, was performed.
Postoperative ophthalmologic examination showed stable findings.

Case 4

In 1986, a 6 year old female presented with moderate proptosis of the left eye and a bluish conjunctival lesion associated with a swelling under the left eye. Visual acuity was 7/10 in the affected eye. Echography showed a cystic lesion with low internal reflectivity and CT- scan of the orbit documented an extraconal retrobulbar mass.
A biopsy was performed and the diagnosis of lymphangioma was confirmed.
In 1987 an increasing size of the tumour was observed and MRI, performed at that time, showed maxillofacial extension. The visual acuity became 3/10 due to a high secondary astigmatism (-6 /170°). Recurrent bleeding within the cheek tumour was documented in 1995. Interferon therapy followed by steroid treatment was given, but without improvement.
The patient was lost for follow-up since 1997.

Case 5

In June 2000, a 29 year old female was referred for acute onset of right proptosis.
A previous diagnosis of ''pseudotumour'' was made in another institute where she had been treated in 1996 without improvement for 6 weeks with oral steroids and systemic non-steroid anti-inflammatory drugs.

Fig 4A. This patient shows a severe proptosis with chemosis.

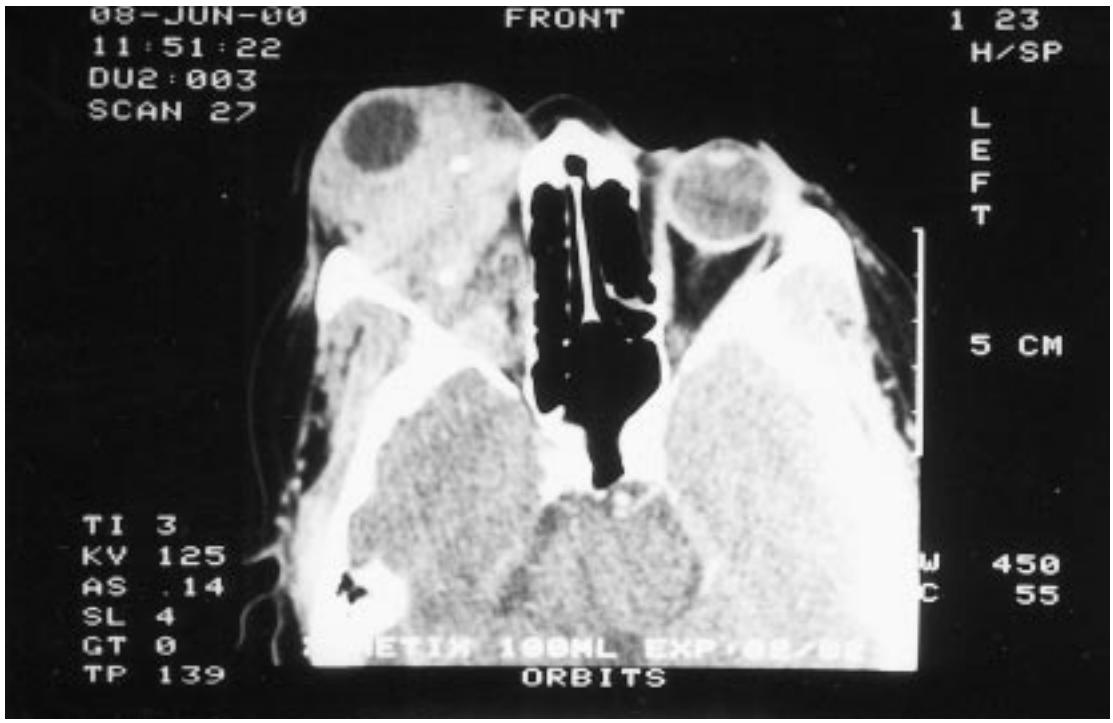

Fig 4B. CT-scan : calcifications are present within the tumour.

Examination on admission revealed a severe proptosis with chemosis, ptosis and restricted ocular motility upward and medially (Fig. 4A). There was no increase in the proptosis with valsalva manœuvre and no bruit on auscultation. The visual acuity was limited to counting fingers in the affected eye with an afferent pupillary defect. Positive findings on slitlamp examination included extreme chemosis, dilated episcleral vessels, high intraocular tension (35 mm Hg by Schiötz tonometry) and manifest right papilledema,.

CT-scan showed a large mass involving almost the entire intra- and extraconal orbit. The optic nerve and orbital muscles were compressed. Multiple areas of calcification were present within the mass and there was a bony orbit enlargement (Fig. 4B). Resection biopsy was performed and pathological findings were suggestive of orbital lymphangioma.

DISCUSSION

Lymphangioma is a rare, diffusely infiltrating benign vascular tumour, most commonly diagnosed in the first or second decade of life (3,11, 15, 17). Orbital involvement may include the lids, conjunctiva or orbit (1,12). There has been no consensus regarding the morphologic nature of lymphangioma and several authors have questioned the validity of the term orbital lymphangioma.

A new terminology ''combined venous lymphatic vascular malformations'' has been proposed. It is believed that the lesions are a variant of venous malformations (6, 7, 8). According to their haemodynamic relationship, division was made into three groups: no flow, venous flow and arterial flow lesions (4, 5, 9, 15). In their survey, Rootman et al. (11) have proposed to classify lymphangioma into superficial, orbital and combined lesions according to the anatomic localization.

Classification of our cases is found in Table 1.

Age at recognition of symptomatic tumour in our series was within the first decade for 3 of 5 patients. All our cases were unilateral as in most reported cases. There is no significant race or sex predilection (6, 15). The preva-

Table 1: *Associated lesions in our 5 reported orbital lymphangiomas*

CASE	1	2	3	4	5
CONJUNCTIVA	-	+	+	+	-
PALATAL	-	+	+	-	-
FACIAL	-	-	-	+	-
COMPRESSIVE OPTIC NEUROPATHY	-	-	-	-	+
INTRACRANIAL VENOUS VASCULAR ANOMALY	-	-	+	-	MRI not performed
CLASSIFICATION	Orbital	Combined	Combined	Combined	Orbital

lence of orbital lymphangioma has been reported to be between 1-3 % of all orbital mass and 10 % of all vasculogenic tumours (12). Ipsilateral extraorbital localization was observed in our cases 2, 3 and 4. In a retrospective study palatal lesions (our cases 2 and 3) are found in 13,8 % of cases and maxillofacial vascular lesions (our case 4) have been reported to be 11,8 % of cases (10). Associated intracranial vascular venous anomaly (our case 3) has recently been reported in 28 % of cases (9), but previous studies showed a much lower incidence (1,9 %) (15,18).

Clinical features, treatment and prognosis are correlated with the location of the lymphangioma.

- The ***superficial lymphangioma*** typically presents a small multilobulated cystic mass of bulbar conjunctiva or/and the lid that neither affect vision nor displace the globe. When cosmetically unacceptable, they can be removed with good results.
- The ***orbital lymphangioma*** is located behind the orbital septum and involves the intraconal or/and extraconal space. Usually they will present with a sudden proptosis and occasionally with pain due to spontaneous intralesional haemorrhage or to lymphoid proliferation in the connective tissue of the tumour during an upper respiratory infection. Other signs may include ptosis, eye movement restriction and compressive optic neuropathy with papilledema and secondary glaucoma.
- The ***combined lymphangioma*** consists of both the superficial and deep form and involves the intraconal, extraconal preseptal and postseptal spaces. It has an anterior component with multiple conjunctival or lid cysts. Similar cystic lesions of mucous membrane of the mouth and the palate are often observed in this group (our cases 2 and 3).
  Spontaneous intratumoral haemorrhage may result in peri-orbital ecchymosis, ptosis and amblyopia. Optic nerve dysfunction may occur as the result of acute expanding tumour. The patient may require surgery either on urgent basis in case of acute retrobulbar haemorrhage with optic nerve compression or on elective basis for cosmetic problem.
- An isolated ***intracranial vascular venous anomaly*** associated with a cavernous haemangioma, as occurred in our patient 3, has been reported previously in patients with combined lymphangioma and related cerebral haemorrhage has been documented (9, 12, 16).

Marked ptosis and high astigmatism may cause deprivation or anisometropic amblyopia (our cases 2 and 4).

Rapidly progressive proptosis in a child will rise suspicion for rhabdomyosarcoma (17) and the need of diagnostic biopsy has to be discussed. In our 5 patients, unlike orbital varices, neither bruit or pulsation, nor variation in the grade of proptosis with valsalva manœuver was present on examination.

To establish the diagnosis, CT-scan and MRI of the orbit are obligatory. CT-scan usually shows a multilobulated cystic mass within the orbit and intralesional calcifications (12) may be demonstrated (our case 5) (Fig. 4B).

MRI is the modality of choice to identify the haemorrhagic cyst as well as the lymphatic cystic component (2, 10).

Haemorrhagic cyst (acute and subacute) shows hypersignal on both MRI-T_1 and -T_2-weighted images (Fig. 1B) whereas lymphatic cysts are hypo-intense on T_1-weighted images and hyperintense on T_2-weighted images (Fig. 1C).

MRI study is most helpful in documenting fluid-fluid level within the multicystic lesion. Pathological examination of a lymphangioma reveals a non-encapsulated lesion with variable sized cystic spaces, lined by flattened endothelial cells. Pericytes and smooth muscles are absent in the vessels wall. This feature, the progression by recurrent intralesional haemorrhages and the absence of clinical response to interferon and corticotherapy (our cases 3 and 4) help to distinguish lymphangioma from cavernous haemangioma.

Treatment options for orbital lymphangioma include conservative management, partial surgical resection of the major cyst, needle aspiration, intralesional injection of sclerosing agents and local radiotherapy (5, 7, 8, 13).

Because lymphangioma is a non-encapsulated tumour, which forms a labyrinthic network of interconnected channels with arborization in the orbital tissue, complete removal is not possible without orbital exenteration (6, 7).

The indication for surgery reported in retrospective studies (1, 3, 11, 15) includes acute orbital haemorrhage with compressive neuropathy, severe pain secondary to intraorbital hypertension, cosmetic problem, amblyopia and diagnostic surgery.

Conservative management has been reported to be followed by spontaneous resolution and normalization of visual acuity in selected cases (1, 7, 10, 17).

Injection of sclerosing agents (sodium tetradecyl sulfate) may be a useful therapeutic option for some patients with eyelid or orbital lymphangioma, especially if no surgery preceeded. More recently the use of intralesional injection of OK-432 has been reported to decrease the volume of the lymphangioma without functional side effects, but further studies are needed to determine long term efficacy and safety (14).

Conjunctival lymphangioma has been treated by ablation with CO_2 laser, cryotherapy and surgical removal (1, 15). Fractionated β-irradiation has been used as an alternative therapy for conjunctival lymphangioma with no further lesion progression at 2 years follow-up. However the low proliferation rate of lymphangioma makes a complete destruction not possible (1).

Needle percutaneous aspiration has been used to drain haemorrhagic fluid but there is a high risk of optic nerve puncture and the recurrence rate is very high (13).

CONCLUSION

Orbital lymphangioma has to be considered in any case of childhood proptosis. Associated extraorbital localizations, particularly intracranial vascular venous anomaly, has to be ruled out. A multidisciplinary approach is needed and the treatment has to be as conservative as possible, if vision is not at risk and cosmetic is acceptable.

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