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 ophthamologique, 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 retrouvées. 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.
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.

Case reports
Case 1
In September 1999, a 5 year old boy presented with a sudden onset of right proptosis (4 mm

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

Fig 1B. MRI-T1 axial: multilobulated intra-orbital mass with internal hyperintense signal representing a recent bleeding.

Fig 1C. MRI-T2 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 T₁ and T₂ 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 lymphangiomma was made on clinical ground and a 6 month follow-up was advised.

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.](image)
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 3C. MRI-T1 after gadolinium: shows the absence of contrast enhancement.

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

Fig 3E. Intracranial developmental vascular anomaly.
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-scans 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.
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 manoeuvre and no bruit on auscultation. The visual acuity was limited to counting fingers in the affected eye with an afferent pupilary 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-
ence of orbital lymphangioma has been report-
ed to be between 1-3 % of all orbital mass and
10 % of all vasculogenic tumours (12). Ipsil-
lateral 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 re-
cently been reported in 28 % of cases (9), but
previous studies showed a much lower inci-
dence (1,9 %) (15,18).

Clinical features, treatment and prognosis are
correlated with the location of the lymphangi-
oma.
- The superficial lymphangioma typically pre-
sents a small multilobulated cystic mass of
bulbar conjunctiva or/and the lid that nei-
ther affect vision nor displace the globe. When
cosmetically unacceptable, they can be re-
moved with good results.
- The orbital lymphangioma is located behind
the orbital septum and involves the intra-
conal or/and extraconal space. Usually they
will present with a sudden proptosis and oc-
casionally with pain due to spontaneous in-
tralesional haemorrhage or to lymphoid pro-
liferation in the connective tissue of the tu-
mour during an upper respiratory infection.
Other signs may include ptosis, eye move-
ment restriction and compressive optic neu-
ropathy with papilledema and secondary glau-
coma.
- The combined lymphangioma consists of both
the superficial and deep form and involves the
intraconal, extraconal preseptal and post-
septal spaces. It has an anterior component
with multiple conjunctival or lid cysts. Sim-
ilar 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 oc-
cur as the result of acute expanding tumour.
The patient may require surgery either on ur-
gentbasis in case of acute retrobulbar haemor-
rhage with optic nerve compression or on
 elective basis for cosmetic problem.
- An isolated intracranial vascular venous anom-
aly associated with a cavernous haemangi-
oma, as occurred in our patient 3, has been
reported previously in patients with combined
lymphangioma and related cerebral haemor-
rhage has been documented (9, 12, 16).

Marked ptosis and high astigmatism may cause
deprivation or anisometric 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, nei-
ther bruit or pulsation, nor variation in the grade
of proptosis with valsalva manoeuver 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 cy-
sic component (2, 10).
Haemorrhagic cyst (acute and subacute) shows
hypersignal on both MRI-T1 and -T2-weighted
images (Fig. 1B) whereas lymphatic cysts are
hypo-intense on T1-weighted images and hy-
perintense on T2-weighted images (Fig. 1C).

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Table 1: Associated lesions in our 5 reported orbital lymphangiomas

Clinical and imaging features of the lymphangioma are described in detail in the ensuing sections.
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 preceded. 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 CO₂ 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.

BIBLIOGRAPHY

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