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Orbital Lymphangioma In A Case Report Of 5 Patients.

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Abstract:
Introduction:
Orbital lymphangiomas are rare histologically benign angiodysplasias with progressive development, usually diagnosed in children between 1 and 15 years of age, with a female predominance. They are often difficult to treat surgically, especially in their diffuse forms.
Materials and methods:
Five cases were operated on, all of them female, with an age ranging from 6 to 27 years, with a duration of evolution varying between 2 and 8 months. Their visual acuity was preserved between 7/10 and 10/10, except in one child where it was 1/10. The lesions were mostly located on the left side (02 cases on the right side, and 03 cases on the left side), only one patient presented a subacute exaggeration of her exophthalmos following an intracystic bleeding.
Results:
The treatment was surgical for all patients, thus ensuring a subtotal removal of the lesions due to their invasive nature. Four lymphangiomas were operated on by the superior trans-palpebral approach (one patient who was initially treated by an external orbitotomy and then reoperated by the superior trans-palpebral approach) and one case by the inferior subciliary approach. The different surgical cures consisted of an ablation of the most voluminous cystic parts at the origin of the symptomatology.
Conclusion:
Orbital lymphangiomas are benign lesions, but serious because of their invasive and hemorrhagic character. Surgical removal is particularly delicate because of the absence of a cleavage plane with respect to healthy tissue. But it can often give a satisfactory aesthetic and functional result
Keywords*: Lymphangioma, Orbital tumor, Orbitotomy, Ophthalmologic approaches, Transpalpebral approach*

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I. INTRODUCTION

Lymphangiomas are benign hamartomatous tumors with low flow rate, usually diagnosed in children [5], with progressive development, most often involving the head and neck. Orbital localization is rare (1 to 2 % of orbi-tal tumors) [7], exophthalmos remains the main symptom, with progressive evolution, sometimes rapid following a bleeding or a local infection

Often affects a young population between 1 and 15 years of age, with a clear female predominance. [5]

Imaging is essential for the diagnosis of which MRI is the reference examination which shows multicystic, infiltrating lesions of intra or extraconical location. Their infiltrative nature with absence of cleavage plane with the surrounding structures excluding a complete surgical resection and their clear tendency to intralesional hemorrhagic recurrence [3]. For this reason, surgery remains a subject of discussion and a conservative attitude, except for complications, is advocated by several authors.

II. MATERIALS AND METHODS

This is a retrospective study of five patients with intraorbital lymphangioma, managed over a period of 07 years from 2015 to December 2022 (which represents 6.41% of all orbital pathology during this period in our department). All these patients had clinical radiological and histological criteria that were in favor of the diagnosis of orbital lymphangioma.

III. RESULTS

-This is a study of five cases taken in our department, all of them female, with an age between 6 and 27 years, with a duration of evolution that varies between 2 and 8 months. The visual acuity of our patients was preserved between 7/10 and 10/10, except for one child in 1/10 (fig. 1-2- 3). Only one patient presented preoperatively a subacute exaggeration of her exophthalmos following an intracystic bleed (figure 2).

All our patients were explored by imaging (CT and cerebro-orbital MRI) which generally showed the appearance of a multicystic mass whose limits are a little clearer on MRI than on CT, often poly-lobed, which can be spontaneously discretely hyperintense on T1 sequences, not enhanced by gadolinium. Lymphatic cysts are hypointense on T1 and hyperintense on T2 sequences (figure 1).

The lesions were mostly located on the left side (02 cases on the right side, and 03 cases on the left side) (fig2, fig3).

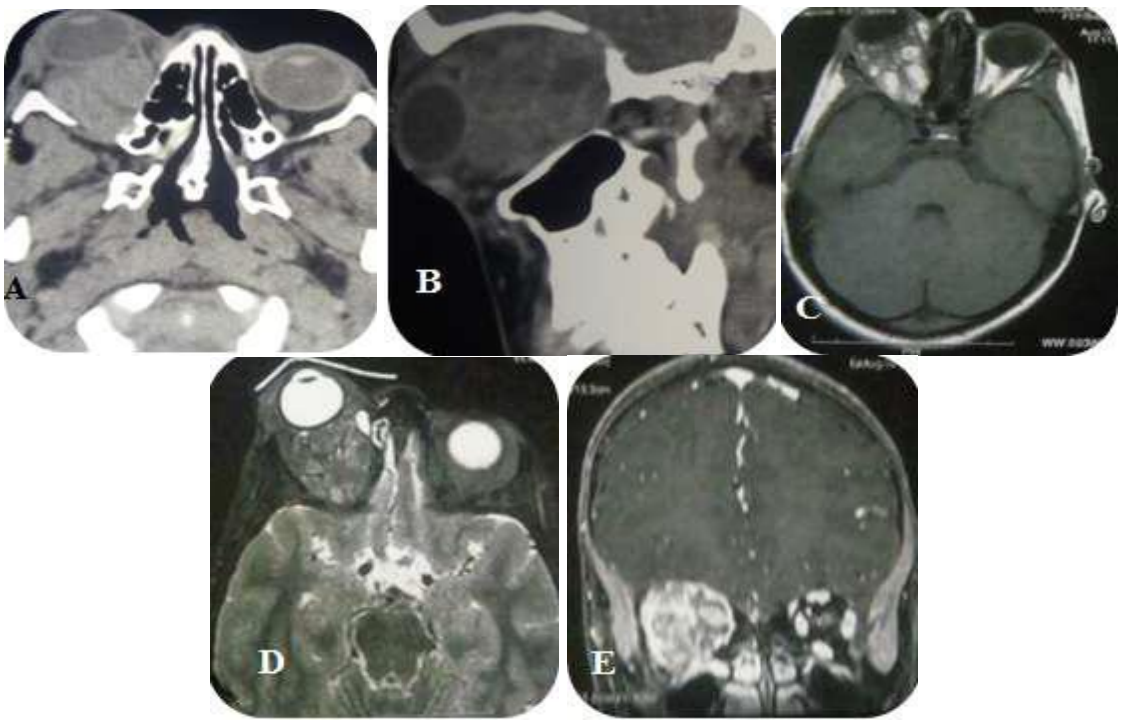


Figure 1: Cerebral CT in axial and sagittal section shows a microcystic infiltrating lesion of the right orbit. Brain MRI in axial and coronal T1 and T2 fat suppression section shows a heterogeneous process with microcystic phleboliths infiltrating the structures of the right orbit.

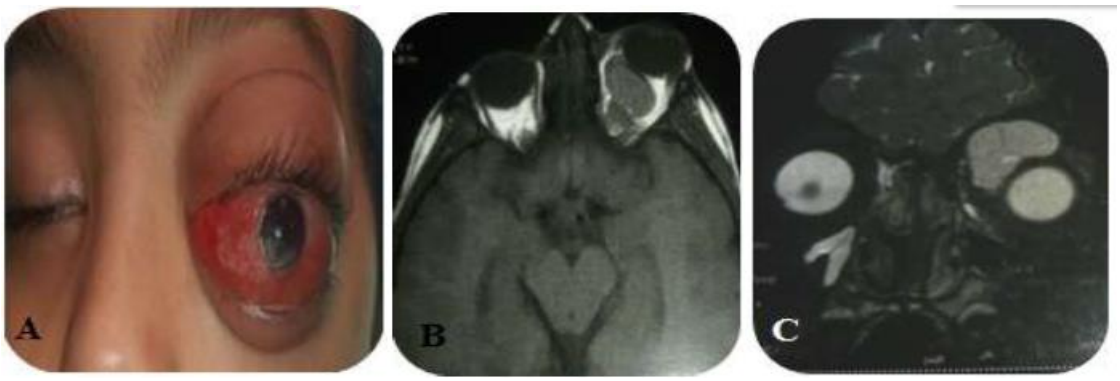


Figure 2: Preoperative clinical appearance of malignant left exophthalmos following intra-tumoral bleeding. Cerebral MRI in axial and coronal section in flare and T2 in fat suppression shows several intra conical cystic logettes filled with injured blood.

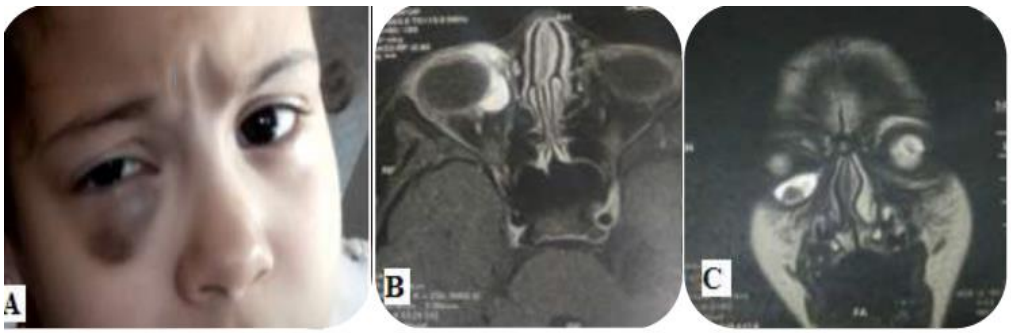


Figure 3: A: preoperative photo of a bluish subcutaneous swelling, B,C:cerebral MRI in axial and coronal section in fat suppression flair show an intra and extraconical cystic lesion.

-The treatment was surgical for all our patients, the majority of which were removed by ophthalmologic approaches.

-Four lymphangiomas were operated on via the superior trans-palpebral approach (Figure 5-6) (one patient who was initially treated with an external orbitotomy and then reoperated on via the superior trans-palpebral approach) and one case was operated on via the inferior subciliary approach (Figure 3). The different surgical cures consisted of a removal of the most voluminous cystic parts at the origin of the symptomatology.

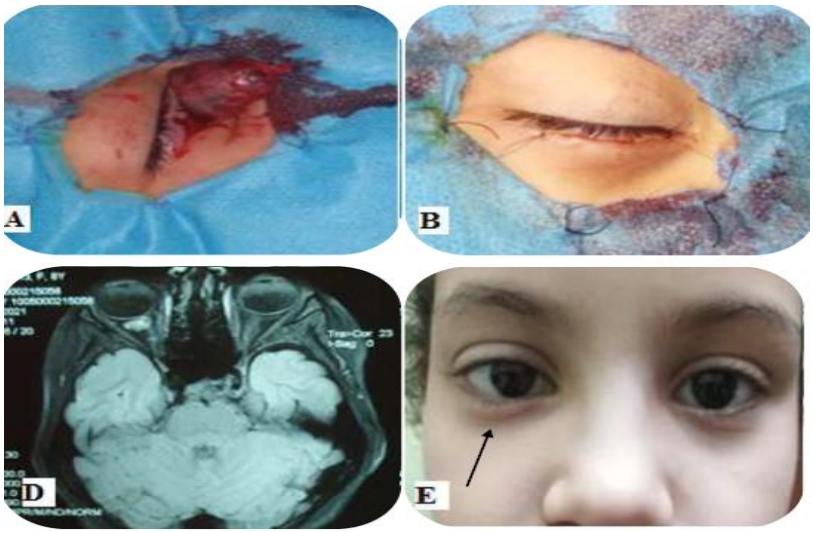
The upper and lower trans-palpebral routes are aesthetic and efficient approaches for intra and extra conical lesions of more or less reduced volume.

- Surgical technique of the superior transpalpebral approach:

The incision is trans-palpebral superior (figure 5) in a natural fold that begins at the level of the supra-orbital notch that ends at the level of the external canthus of the eye, then section of the orbicularis muscle of the eye, thus exposing the anterior face of the levator muscle of the upper eyelid, We then move towards the insertion of the orbital septum on the anterior face of the levator muscle as far as the bony edge, then incision of the periosteum (be careful with the supraorbital nerve in its notch), which gives access to the intraorbital contents and the lesion.

This approach can expose the external pillar of the orbit, which can be removed, which can optimize the exposure and give an interesting variant of the kronlein approach.

*the trans-palpebral or sub-inferior ciliary approach (figure 4) is interesting for lesions located in the inferior quadrant of the orbit, either intra- or extra-conical



<u>Figure 4</u>: A: Perioperative photograph of a cystic lymphangioma under the lower palpebral region, B: : skin closure showing the subciliary incision, C: control brain MRI shows a tumor remnant in the retro-bulbar region,D: postoperative photo of the girl

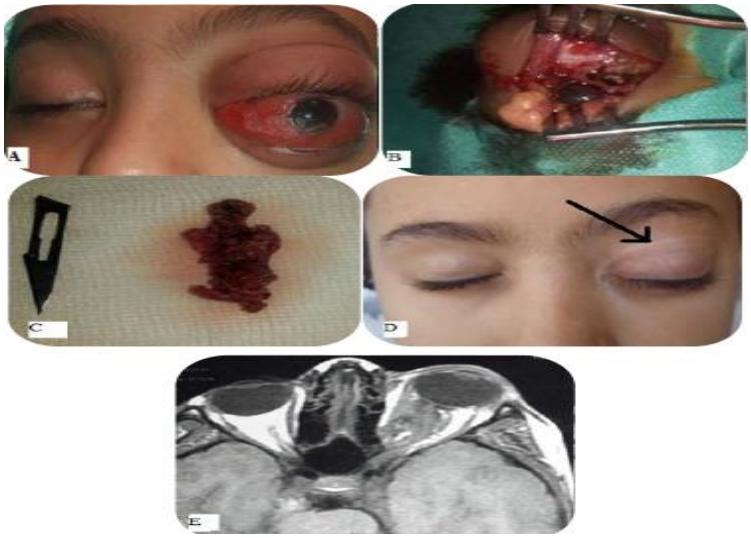


Figure 5: Left orbital lymphangioma. A: Photo of an acute grade III exophthalmos, it also shows the line of the upper trans-palpebral skin incision, B: photo perioperative showing the cystic lesion, C: surgical specimen of the cystic part that has bled, D: postoperative photo showing a clear reduction of the exophthalmos: postoperative control axial slice brain MRI shows a reduction of the tumor volume.

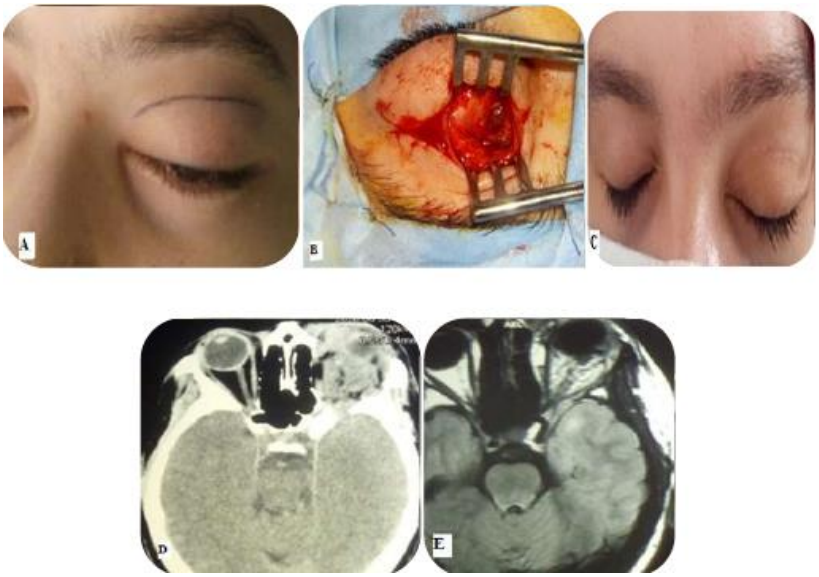


Figure 6: photos of surgery of a multicystic lymphangioma. A: photo of the incision line B: photos of a preoperative aspect of a cyst composing the superior trans-palpebral lymphangioma, C: aesthetic result of skin closure, D: control brain CT and E :Control brain MRI in T2 axial slice showing a reduction of the tumor volume.

IV. DISCUSSION

Lymphangiomas are rare histologically benign congenital angiodysplasias with progressive development and hemodynamically inactive (slow flow vascular malformation) [5], frequently localized in the head and neck (20% in the orbit). And represent about 2% of orbital tumors) [7].

The orbit is normally devoid of lymphatic structure, paradoxically, lymphangioma can reach the orbit, so the vascular tumoral nature seems most likely, Wright suggests that lymphangiomas are venous malformations, however clinical hemodynamic and histological studies confirm that they are hamartomas, with

20% of orbital location. [5]. The growth of this malformation is caused by repetitive hemorrhage and not by cell growth [8]

Preferentially affects the female sex up to 77% in some studies [5] and is 100% in our series.

Clinically, they are manifested by an exophthalmos of progressive installation, whether axial or not, which can evolve through remission attacks of variable reversibility (these attacks are often preceded by ENT infections or local trauma) and the valsalva maneuver does not aggravate this exophthalmos, which is what makes the difference with the other venous malformations

This major sign is associated with dystopia, limitation of eye movements and tissue infiltration (frog's feet in case of eyelid infiltration), periorbital pain and ptosis are frequent [5], while diplopia is rare.

Sometimes the mode of revelation is acute following an intracystic bleeding, causing a brutal exophthalmos and a decrease of the visual anuitae [7], one case in our series.

The intraorbital localization is variable rarely limited to an intraconical and extraconical compartment and even extraorbital.

The diagnosis is made by imaging which is essential to confirm it. Echo-Doppler shows a cystic tumor with slow flow. MRI remains the examination of choice which shows the appearance of a heterogeneous, poly-lobed tumor, with a liquid signal of variable intensity related to hemorrhagic attacks of different ages, it allows to distinguish macrocystic (> 1 cm) and microcystic forms, and to search for a possible extraorbital extension. The presence of phleboliths points to a veno-lymphatic origin.

These lesions are often difficult to treat surgically, especially in their diffuse forms. [4].

Surgery is therefore often limited to a limited exeresis, which is still a subject of discussion [2]. The attitude currently recommended is mainly conservative. Indeed, these are very infiltrating lesions for which total surgical cleavage is impossible [photos]. Surgery is reserved for the most voluminous lesions, responsible for important exophthalmos with functional threat [4].

The approach depends on the location and size of the tumor, with patients who are not very symptomatic simply being monitored [5].

In some studies [2], initial systemic corticosteroid therapy was initiated to reduce the tumor volume, and then a Kronlein-type orbitotomy was adopted with complete resection of the lesion.

Progressive flare-ups, even important ones, should be treated medically as a first step [5].

The indication for surgery in our series was based on: an aggravation of the symptomatology following this malignant exophthalmos, and in the other cases the aim was aesthetic.

However, with or without surgery the probability and time of onset of bleeding remains unpredictable [2].

V. CONCLUSION

Orbital lymphangiomas are benign lesions, but serious because of their invasive and hemorrhagic character. Surgical removal is particularly delicate because of the absence of a cleavage plane with respect to healthy tissue. But it can often give a satisfactory aesthetic and functional result

Declaration of interest: The authors declare that they have no interest.

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