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Cavernous Intraorbital Hemangiomas: About 16 Cases.

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SUMMARY:
Introduction :
Intra-orbital cavernous hemangiomas (or angiomas) are the most common benign orbital tumors in adults, most often occurring between the 3rd and 5th decade. Of well-defined forms , they are formed by the benign proliferation of dilated capillaries. The evolution is slowly progressive, often causing exophthalmos which may be accompanied by a decrease in visual acuity or ocular motility disorder . Seat often intra conical. Their treatment of choice is surgery.
Materials and methods :
We bring a series of sixteen cases, supported, during a period of 05 years, whose age varies between 20 to 63 years for adults and 2 years for the only child. The evolution varies between one year and 12 years, with an average of 6.5 years. We have a clear female predominance (13 women). The seat was on the right for six patients, and 10 cases on the left side.
One case underwent blindness surgery, two had light perception on clinical examination, the other patients had preserved visual acuity (between (5/10 and 10/10).
Results :
The treatment was surgical for all our patients, thus ensuring total excision without morbidity. six cases operated on via an external orbitotomy via a fronto -orbital approach and two cases via the same route associated with removal of the external pillar, three cases operated on via the trans-conjunctival route with ophthalmologist surgeons, one case via the sub-brow route, and three via the upper trans - palpebral route (two of which were enlarged by removal of the external pillar of the orbit), and three cases via the lower subciliary route.
Conclusion
The cavernous hemangioma is slowly growing tumors which is the most common primary benign lesion of the orbit. The clinical picture is stereotyped. The treatment of choice is surgery, generally guaranteeing satisfactory anatomical and functional results.
KEY WORDS: Cavernous hemangioma, vascular tumor, orbital approches, Orbitotomy, Trans - conjunctival route .

I. INTRODUCTION:

Vascular orbital tumors are most often benign, the most common are cavernous hemangiomas. Represent 3 to 14% of all orbital tumors according to the main published series [6], and 50 to 80% of orbital vascular tumors [2], often affects young adults, rarely before the age of 20, with a clear female preponderance. The sex ratio can be seven out of ten women. They are often manifested by an axile proptosis, with acquired hyperopia and chorioretinal folds at the fundus [4].

It is a histological point of view of a hamartoma , which hemodynamically behaves like a low-flow arteriovenous malformation , but of a benign nature [3]. Of slow evolution, is most often located intraconically. It often leads to axial proptosis.

Imaging shows a well-defined and encapsulated lesion, hyperdense in iso-hypo signal on T1, a hyper signal on T2, and which is slightly enhanced after injection of contrast products.

Its treatment is surgery which must be done block [6] , especially if the visual function is threatened.

The approach is based on the seat and volume of the tumor, in fact surgery has evolved since Kronlein who described his approach first by lateral orbitotomy in 1889, which will be modified by several authors, then other approaches have been developed such as the neurosurgical approach, the anterior cutaneous or trans -conjunctival ophthalmological approaches, and the endoscopic approaches giving significant advantages.

II. MATERIALS AND METHODS:

The study is retrospective, on a series of sixteen cases with intra-orbital hemangiomas, treated over a period of 07 years (from June 2015 to December 2022). All these patients had the radiological and histological clinical criteria which were in favor of the diagnosis of cavernous hemangioma.

III. RESULTS:

It was about a series of 16 patients whose age varies between 20 to 63 years for the adults and 2 years for the only child with an average of 33

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years. The evolution varies between one year and 12 years, with an average of 6.5 years. We had a clear female predominance (13 women). Clinically, proptosis was unilateral axile, found in 13 cases (fig1).

One patient was blind preoperatively, two had light perception, the other patients had visual acuity varying between 5/10 and 10/10.

Figure 1: Preoperative appearance A, B of a left cavernous hemangioma (Naphzeiger test). Cerebral MRI C, D in axial and coronal section in T2, shows the appearance of a cavernous hemangioma in the form of a homogeneous intraconical tissue process, well limited in flaire iso signal and hyper signal T2, gradually enhancing.

cerebro -orbital MRI) which generally showed well-defined, homogeneous encapsulated lesions which slightly enhanced after injection of contrast products. On T1-weighted MRI, the lesions are isointense in relation to the muscles, the contrast enhancement may show an irregular appearance like a sheet of fog or an apple tree of flowers. (fig2)

The location of the lesions was on the right for six patients, and on the left for 10 cases. The hemangiomas were intra-conical in 13 cases, including one at the level of the orbital apex (fig3), and two cases extra-conical (fig4).

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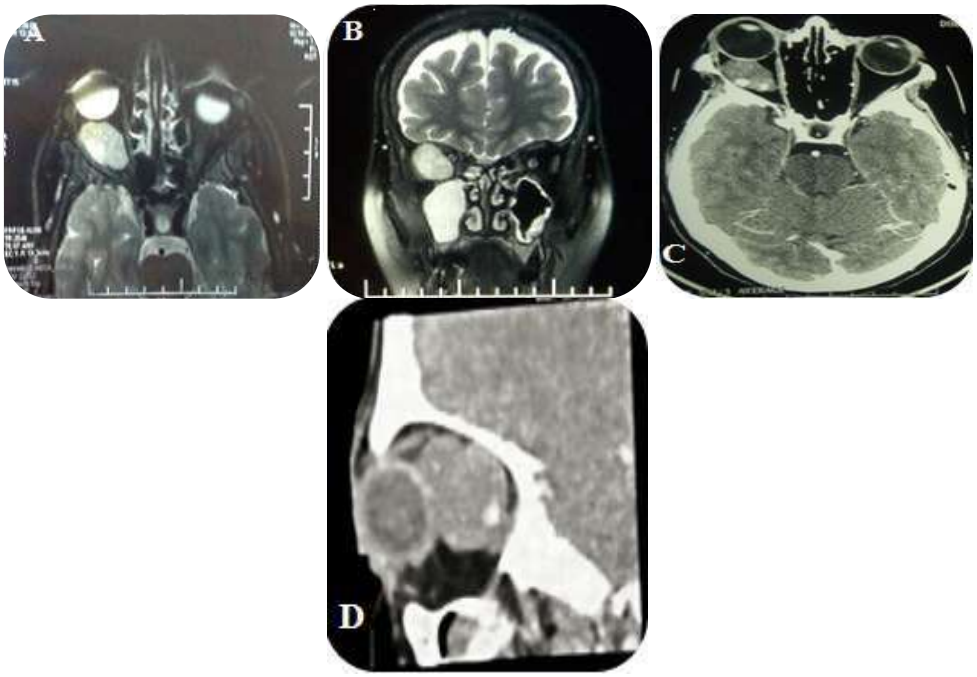

Figure 2: Cerebral MRI A, B in axial and coronal section in T2 shows a hemangioma intraconical cavernous in well-limited homogeneous T2 hypersignal. Brain CT. C, D, in sagittal and coronal axial section shows a dense process from the outset

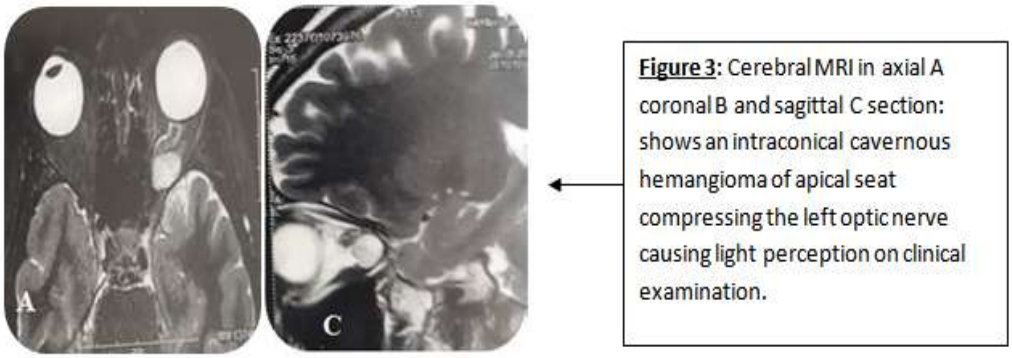

Figure 4: Cerebral MRI in axial and coronal section in T2 shows an extra conical cavernous hemangioma

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-The treatment was surgical for all our patients thus ensuring a total excision, we do not note any morbidities except for one patient who was already blind.

Six cases operated by an external orbitotomy by a fronto -temporal approach, two of which by the same route associated with removal of the external pillar, three cases by upper trans -palpebral approach (including two enlarged by removal of the external pillar of the orbit) , three patients operated by trans -conjunctival route in collaboration with ophthalmologist surgeons, three cases by a lower subciliary route, and one case by sub-brow way.

To simplify, three operative techniques have been used.

1- **Lateral approach:** by performing a **Kronlein variant** either by a fronto -temporal neurosurgical incision then removal of the external pillar and the lateral wall of the orbit (fig 5) (for aesthetic reasons, the incision is hidden at the level of the scalp), or by an anterior superior trans -palpebral approach (fig 6) over its entire length, which is widened towards the external pillar of the orbit, which will be deposited thanks to the oscillating saw, also taking away part of the lateral wall. The orbital contents were then exposed after opening the periorbit. At the end of the excision, the flap was repositioned and held in place with resorbable suture.

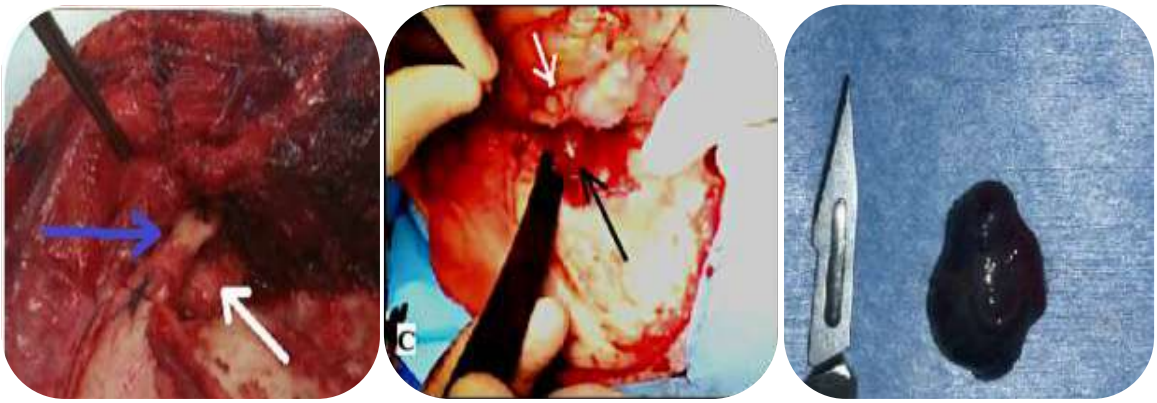

Figure 5: External orbitotomy through a right fronto-temporal skin flap. A: intraoperative photo of a right lateral orbitotomy white arrow, intraoperative photo of excision of the haemangioma in black the white arrow indicates the orbit after removal of the external pillar of the orbit, D: surgical specimen of a haemangioma cavernous.

Figure 6: A: intraoperative photo showing the upper trans-palpebral skin incision, B: removal of the external pillar of the orbit, C: surgical specimen of a cavernous hemangioma, D: CT scan cerebral reconstruction in 3D after a few months shows the consolidation of the external pillar, E: postoperative photo showing the aesthetic scar in a palpebral fold. F: cerebral CT of control in axial section is satisfactory.

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Figure 7: A: preoperative photo of the lower ciliary skin incision, B: photo surgical excision of a cavernous hemangioma, C: postoperative result, E: CT scan cerebral control in axial section does not show recurrences.

2- Anterior transcutaneous approach For tumors located in the upper or lower part of the orbit, these are aesthetic and effective approaches for intra and extra conical lesions of reduced volume. The incision was made at the level of the upper palpebral fold (fig6), or under the lower ciliary (fig 7) depending on the seat of the lesion, The orbicularis muscle then the orbital septum will then be incised in turn, the dissection is done in a intraconical fatty plane along certain corridors between the muscles which will be retracted so as to expose the tumor mass, thus facilitating its dissection which must be delicate according to its cleavage plane.

3- Trans -conjunctival route : carried out in collaboration with ophthalmologists, this is a very interesting route in intraconical orbital pathologies of reduced volume of the anterior and/or medial seat. The incision was made at the level of the insertion of the oculomotor muscles, which will be individualized on tractor wires, a lateral canthotomy (could be carried out in order to optimize exposure), then a gentle para-ocular dissection was carried out until retro-bulbar by soft objects, possibly under operative microscopy, and the lesion will be delicately mobilized and dissected from these different liners that surround it.

Figure8: right trans-conjunctival pathway 1: preoperative photo showing exposure and preparation of the juxta corneal conjunctiva. 2: photos showing the incision of the pericorneal conjunctiva. mooring of the rectus muscles by suspension wires. cut of the external rectus muscle then displacement of the eyeball allows intraconical access to the lesion. 3: excision of a cavernous hemangioma.

In our series, no recurrence was observed, and the outcome was favourable, with a reduction in proptosis in all patients, and an improvement in visual acuity, except in the case who was blind.

IV. DISCUSSION:

Cavernous hemangioma is the most frequent vascular tumor of the orbit in adults (80% of them according to the literature) [2], and which varies between 10-15% of orbital lesions [1]. Arises from the benign proliferation of dilated capillaries, which are delimited by fibrous septa,

hence the name cavernoma , giving the macroscopic appearance of a very well-defined, encapsulated mass containing dark red cells [3]. Unlike capillary hemangiomas, they have no tendency to spontaneous regression.

Our series from the Cherchell hospital includes sixteen patients, which represents 18% of operated orbital lesions (B.Yakoubi in his series finds a rate of 16%, in the Cophignon summer series of 10.15%). Often affect young adults with an average age of 41 years in our study, yan j et al report an average age of 39 years [5] . Harris and Jakobiec [3] out of 66 cases, the mean age of the patients was 42 years.

Regarding the predominance of sex, we had a clear female predominance with a rate of 81%, which is consistent with most of the series whose rate varies between 57% to 91.2%, with a clear female predominance at 91.2%, (Kiratli [5] reports a male predominance of 63%)

The attack was unilateral for all our patients. We find 33.3% of the lesions on the right side and 66.6% of the lesions on the left side, for Cophignion it is 63.3% on the right, 34% on the left [3].

The intraconical seat is 85.71%, and extraconical in 14.28%, comparable with the series in the literature ranging from 100% intraconical up to 75%.

Clinically, hemangiomas are manifested by progressive proptosis [6] axile or not depending on the intra- or extra-conical tumor location , nonpulsatile (72% of cases), painless, except in the event of a complication (inflammatory, hemorrhagic or thrombotic). , and sometimes visual acuity, and ocular motility may be affected.

Three cases had light perception, and one case went blind (6.25), the other patients had visual acuity maintained between (5/10 and 10/10). The mode of discovery can in some cases be fortuitous [4]

Imaging is essential to clarify the diagnosis. In first intention, an orbito - cerebral scanner can confirm the presence of an intra-orbital lesion, to specify certain characteristics (which would be in favor of a malignant lesion) [9]. In the case where the scanner does not allow a conclusion, the MRI and/or the Doppler ultrasound will be useful in order to eliminate the differential diagnoses (hemangiopericytoma, lymphangioma, schwannoma, fibrous histiocytoma, orbital varices) , and will confirm in particular the low vascular flow of the lesion. In our series, the hemangiomas were mainly intra-conical in 87.5%.

On the therapeutic level Only surgery is effective [3]. Surgical excision of the tumor and its capsule must be total. Recurrences are exceptional without malignant transformation. The choice of the optimal approach in the treatment of cavernous hemangiomas of the orbit is essentially dictated by the location and volume of the tumour. The anatomical complexity explains the multiplicity of approaches that have successively been proposed and the need for the intervention of a multidisciplinary team of ENT, ophthalmologists, and sometimes neurosurgeons [6], the neurosurgical approaches treat the lesions located in the posterior two-thirds of the orbit, and the anterior pathways treat those located in the anterior third. All the authors advise to intervene only if the appearance of the first clinical signs (headache, compression of the optic nerve, exophthalmos, diplopia)[2]

The therapeutic abstention can be proposed if the discovery is fortuitous, while remembering the benign character of the tumor and its slow evolution.

The lateral orbitotomy initially described by Kronlein in 1889, is the most classic technique which will later be modified and miniaturized. It makes it possible to approach all intraconical retrobulbar hemangiomas, they represent 37.5% in our series.

Concerning the anterior cutaneous approach (incision at the palpebral or subciliary fold), it is reserved for anterior extraconic tumors, which represents 43.75% in our series, having the advantage of a reduction in operating time and better healing.

The anterior orbitotomy " trans - conjunctival or palpebral route" is indicated in lesions anterior to the equator of the globe, it is increasingly used at the expense of the lateral route with removal of the lateral wall of the orbit which is considered invasive, they represent 18.75% in our study.

Other techniques are also used and described in the literature, such as the use of a gamma knife [2] or cryo excision [7]. Extraction by endonasal endoscopy has shown its value in recent years in apex hemangiomas [4].

Finally, whatever the surgical technique proposed, the prognosis is often excellent and the results are satisfactory in most series.

Recurrences are exceptional and the risk of malignant transformation is nil. Any nodular orbital tumoral lesion of progressive appearance in a middle-aged woman should give priority to the diagnosis of cavernous hemangioma.

V. CONCLUSION:

The cavernous hemangioma is slowly growing tumors which is the most common primary benign lesion of the orbit. The clinical picture is stereotyped. The treatment of choice is surgery, generally guaranteeing satisfactory anatomical and functional results.

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