

PRE SURGICAL EMBOLIZATION OF RECURRENT ORBITAL HEMANGIOPERICYTOMA WITH GLUE

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ABSTRACT

Hemangiopericytoma's are a type of vascular tumors originating from pericyte cells in the eye and they constitute only 3% of all primary orbital tumors.¹ While mostly acting as benign, these tumors have a high recurrence rate in the case of incomplete excision.^{2,3,4} While a reasonable number of cases of hemangiopericytoma have been reported in literature none of them discuss pre-operative embolization and how it improves success rate in surgery.^{6,7,8} The treatment of choice for such tumors is complete surgical excision however due to the vascular nature of the tumors, it is not always achieved. Incomplete excisions come with a number of risks such as increased risk of recurrence and metastasis of the tumor.

The use of n-butyl cyanoacrylate (n-BCA) in pre-surgical embolization of feeding vessel in controlling recurrent orbital hemangiopericytoma is rare. We are therefore reporting a case of pre-surgical embolization in orbital hemangiopericytoma, that recurred for the sixth time after multiple incomplete excisions.

Key words: Hemangiopericytoma; Embolization; n-BCA; Adjunct Therapy

Case Report

A 22 year old woman from a rural area, house servant presented with a sixth recurrence of orbital hemangiopericytoma, 4 years following radical excision of the tumor. Facial appearance at initial examination reveals a protruding mass arising from right lower eyelid and covering whole eye with marked strabismus. The mass was mobile, on compressible and the skin overlying was normal and pinch able (Fig. 1). Her visual acuity was 6/6 in left eye and her acuity was 6/6 in her right eye as well, since her fifth recurrence which was managed with radical excision and enucleation right eye. Her symptoms started 8 years back with swelling upper eyelid after trivial trauma along with discomfort. Some of her symptoms that are associated with hemangiopericytoma were extra-ocular motility limitations with diplopia, displacement of globe and vessel congestion in the conjunctiva.



Figure 1: Pre-Operative Clinical Photograph demonstrating the orbital swelling.

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Patient history found no past neurological problems, headaches or seizures.

Investigations:

We use radiology in delineating the morphology of the tumor prior to any intervention strategy. CT orbit was performed, showing a well circumscribed infra-orbital mass which intensely enhances with contrast. The findings were also consistent with a highly vascular tumor, either orbital hemangioma or hemangiopericytoma. The vascular nature of the tumor and the voids in signal flow that were observed in the MRI helped conclude this was a hemangiopericytoma. After discussion with the patient and internal deliberation, our team decided to perform a cerebral angiography to embolize the tumor pre-operatively. A diagnostic cerebral angiogram showed hyper vascular orbital tumor with multiple feeders coming from the nasal-frontal branch of the right internal maxillary artery. (Fig. 2).

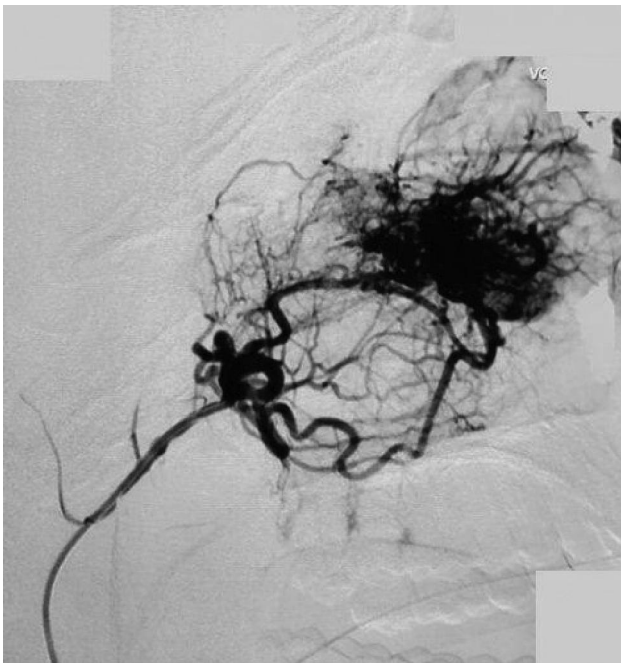


Figure 2A: Selective catheterization and angiography of internal maxillary artery. The tumor appears to be highly vascular with multiple feeder vessels raising from internal maxillary artery.

Differential Diagnosis:

Making the differential diagnosis for orbital hemangiopericytoma, we considered fibrous histiocytoma, rare fibroblastic tumors, nerve sheath tumors, meningioma, synovial sarcoma and solitary fibrous tumor (SFT).

Previously hemangiopericytoma have been misdiagnosed because of similar clinical presentation and microscopic picture. Recently advanced immunohistochemically techniques have facilitated a clear distinction. Differentiation of hemangiopericytoma from SFT is mandatory because hemangiopericytoma is highly aggressive tumor while SFT is benign lesion. Hemangiopericytoma shows inconsistent and weak positivity to CD34, however, SFT shows strong and consistent positivity to CD34.^{9,10}

Treatment:

Considering the extensive vascularity, the tumor was pre-surgically embolized using 1.2 Fr Progreat micro catheter selectively placed distal to feeder. The embolizing agent used was n-BCA diluted in ethane-ethane-diol oil. This dilution was to facilitate distal migration of the n-BCA so as to avoid distal reflux of the agent. 5% dextrose water was concomitantly infused in the proximal guide catheter (Fig. 2B). The tumor was completely and successfully devascularized as a result of the embolization. There was no fear of compromise of central retinal artery as enucleation right eye was already done in previous surgery.



B) Super selective angiogram through 1.2 French micro catheter, n-BCA was injected few mm away from main trunk, there is no glue reflux proximal to main trunk.

Outcomes and Follow-Up:

Pre surgical embolization of orbital hemangiopericytoma was followed by radical excision of tumor under general anesthesia.

Intraoperatively the tumor appeared reddish, hyper vascular and fibrotic.

Estimated blood loss was 1 liter intra-operatively. The patient was discharged in satisfactory conditions 3rd postoperative day. (Fig. 3).



Figure 3: Post-operative image showing no signs of recurrence.

Pathological analysis of the tumor demonstrated highly vascular tumor with variable stromal cellularity. In most areas, neoplasm was composed of proliferation of numerous thick and thin walled stag-horn pattern blood vessels against non-fibrillary background. These cells had round and oval nuclei, eosinophilic cytoplasm and indistinct cytoplasmic borders, diagnosis was spindle cell tumor. Immunohistochemically staining for CD34 was weakly positive. With these morphological and immunohistochemically findings, the diagnosis of hemangiopericytoma was made. Since the patient presented with sixth recurrence, decision of radiotherapy was made and she was being subjected to radiotherapy. Six months post operatively and post radiotherapy the patient is doing well with no signs of recurrence.

Discussion

The first description of Hemangiopericytoma (HPC) was done by Stou and Murray in 1942, who found this to be a vascular tumor of mesenchymal origin.¹¹ It was considered a controversial tumor and its classification has been debated. Latest studies now classify it as a lesion while previous classifications placed, hemangiopericytoma, as a cellular phase of fibrous SFT.¹² SFT/HCP find their origins in Zimmerman pericytes, which are small single cells located in capillaries and post capillary venules. The tumor is considered highly rare making up only 2.5% of soft tissue sarcomas.¹³

"In 2013, the world health organization reclassified, extra pleural solitary fibrous tumors to be a "ubiquitous" mesenchymal tumor of fibroblastic type and rescinded the term "hemangiopericytoma" (HCP)".¹⁴

There is some ambiguity regarding the management of these lesions and only a few reports regarding its clinical management have been published since HPC is a very rare kind of tumor. However, total resection is a definitive treatment to reduce likelihood of recurrence, malignant transformation or need for reoperation.^{15,16}

Intraoperative hemorrhage is a concern in these cases,^{17,18} some authors have suggested pre surgical embolization to reduce the risk of intra-operative bleeding. As a vascular tumor HPC is amenable to trans arterial embolization which has also been suggested to reduce tumor size prior to excision.^{19,20} Methods of embolization have included polyvinyl alcohol, onyx and particulate embospheres (150-300um).^{21,22,23} Couch et al,²⁴ discussed 4 cases of orbital varices that were treated with pre-operative percutaneous injection of n-BCA to embolize the varicosities prior to resection. In our case, we used n-BCA, to occlude the internal maxillary artery. Once the vascular supply of tumor is cut off, significant blood flow was reduced by over 90% in our case. Surgery should be performed 24-48 hours after embolization. A careful visualization of post procedural angiograms can help deciding recession by seeing a visual decrease in the tumor blush.

The risk associated with embolization is inadvertent embolization of central retinal artery. This risk should always be weighed against the benefit of minimizing

intraoperative blood loss.

Radiotherapy when added adjacently, lesions may not be completely excised or in advance and recurrent cases as palliative treatment.²⁵ This high recurrence rate may take place years after the initial surgery and long term follow ups are highly recommended. This makes ours an extra ordinary case of a very advanced hemangiopericytoma, which was completely removed with minimal blood loss after pre-operative embolization with n-BCA glue with the proximal dextrose push technique.

Conclusion

Solitary fibrous tumors or Hemangiopericytoma's are uncommon orbital neoplasm's that present in advanced manners with high risk of intraoperative bleeding and malignant transformation. The bleeding can be managed effectively by pre-surgical embolization with n-BCA followed by a complete surgical excision.

Key Learnings:

- The treatment of orbital hemangiopericytoma is surgery however pre-surgical embolization can be done as an adjunct therapy to improve surgical outcome.
- Pre-Surgical embolization can radically improve chances of recession and recovery.
- n-BCA as an embolic agent is highly effective in embolizing vascular supply of highly vascular hemangiopericytoma.

Conflict of Interest: None

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