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Orbital Angiomyxoma: Bridging Diagnostic Gaps and Enhancing Surgical Outcomes

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ABSTRACT

Orbital angiomyxoma, though rare, presents with gradual proptosis and associated ocular symptoms. Early diagnosis through imaging and histopathology, followed by precise surgical intervention, can significantly improve outcomes, reducing proptosis and ensuring recovery with appropriate follow-up care.

1 | Introduction

Aggressive angiomyxoma is a rare, locally aggressive myxoid mesenchymal neoplasm originating commonly from the soft tissue of the pelvis and perineum of adults comprising both myxoid and vascular tissue [1]. It was first described in 1983, with less than 250 cases having been reported approximately to date, and orbital involvement is a much rarer entity, with only 12 cases reported to the publishing of this case [2]. Being locally aggressive, with few cases of metastasis, it is difficult to diagnose pre-operatively due to its non-specific signs and symptoms [3]. Commonly, this condition is painless, but in our case, the patient did complain of pain. Surgical excision is the primary mode of management, while non-surgical management like hormonal manipulation, radiotherapy, arterial embolization, etc. has also been carried out with variable success rates [3]. The aim of this case is to show how this rare tumor presented to us and how we dealt with it in a resource-limited settings like ours.

2 | Case History/Examinations

We report a case of a 54-year-old male patient who presented to the ophthalmology OPD with complaints of right eye swelling for the past 6 months and pain in the right eye for 1 year. The pain was insidious in onset, dull in nature, and mild in severity, with no radiation or aggravating factors. It was relieved by medication. The swelling began 6 months ago, gradually increasing in size. On examination, the patient's systemic findings were within normal limits. Ocular examination revealed that bilateral visual acuity was 6/6, with a normal head posture. The eyelids were normal in appearance bilaterally. On further examination, there was proptosis of the right eye, which was non-axial, with inferior displacement. On palpation, an orbital mass was identified, which was ill-defined, non-tender, soft in consistency, with a smooth surface involving the superior orbit. The lateral aspect of the orbit was free, and the mass was non-adherent to the overlying skin, with no induration or discharge. The size of the mass was measured as 1.5 cm × 1 cm, and there were no pulsations or thrills. The posterior extension of the mass could not be reached, and resistance was noted on retropulsion. No significant retro-orbital changes were appreciated, and extraocular movements were equal in all meridians.

On slit-lamp examination, the conjunctiva of both eyes was normal, with a clear cornea and brown iris showing a normal pattern. The anterior chambers of both eyes were of normal depth and quiet, and the pupils were round and regular, measuring 3–4 mm

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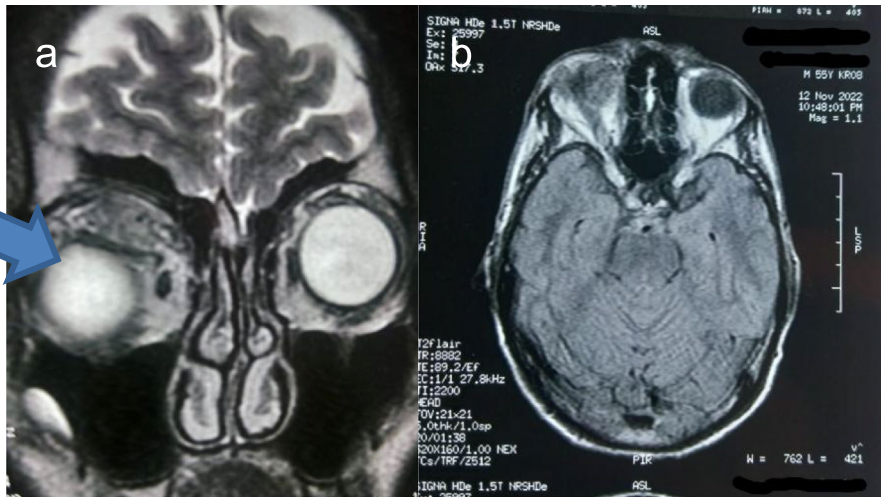

FIGURE 1 | MRI orbit coronal (a) and axial (b) view.

in size, with both direct and consensual pupillary reactions intact. On fundus examination, the optic disc was pink, round with sharp margins, and a cup-to-disc ratio of 0.2:1, with a healthy macula and good foveal reflex. Goldman applanation tonometry revealed an intraocular pressure of 12 mmHg in both eyes. On Luedde's exophthalmometer, proptosis of the right eye was found to be 15 mm. Following thorough ocular examination, the patient was advised to undergo further imaging and investigations.

3 | Methods (Differential Diagnosis, Investigations, and Treatment)

Differential diagnoses considered for the patient's condition included orbital tumors, inflammatory lesions, and other causes of proptosis such as thyroid eye disease or infections. The patient underwent an MRI of the orbit, which revealed an ill-defined heterogeneous intensity lesion at the supero-medial aspect of the right orbit, extraconally, producing proptosis. Heterogeneous signal changes were noted involving the superior rectus muscle and adjacent regions with enhancement (Figure 1). There was a pressure effect on the right ocular bulb, and the findings were suggestive of a mesenchymal mass. Following these findings, the patient was advised to undergo surgical intervention. A right-eye superior orbitotomy with an upper lid split approach was planned. Intraoperatively, a globular, firm, well-encapsulated reddish mass was identified, measuring 3 cm × 1.6 cm × 1.3 cm, which was carefully excised and sent for histopathological examination. Postoperatively, the patient was discharged on antibiotics and corticosteroids and was asked to follow up in 2 weeks with the histopathology report.

4 | Results and Conclusions (Outcomes and Follow-Up)

The histopathological examination revealed an encapsulated tissue with a multi-lobular arrangement of spindled stromal cells and a prominent vascular component set within a copious matrix (Figure 2). The stroma was relatively hypocellular, and the vessels ranged from medium to large size, evenly distributed throughout. The spindle cells showed delicate eosinophilic bipolar cells with oval to round nuclei, with a few cells exhibiting binucleation to multinucleation. The vessel walls showed hyalinization with perivascular concentric condensation of collagen fibers and perivascular lymphocytic infiltrates. Focal areas displayed lymphocytic aggregates with mixed inflammatory cell infiltrates, predominantly composed of neutrophils, lymphocytes, plasma cells, and occasional eosinophils, embedded in a dense fibro-collagenous stroma. Based on these findings, the final diagnosis was consistent with an inflammatory orbital mass, likely a benign mesenchymal tumor.

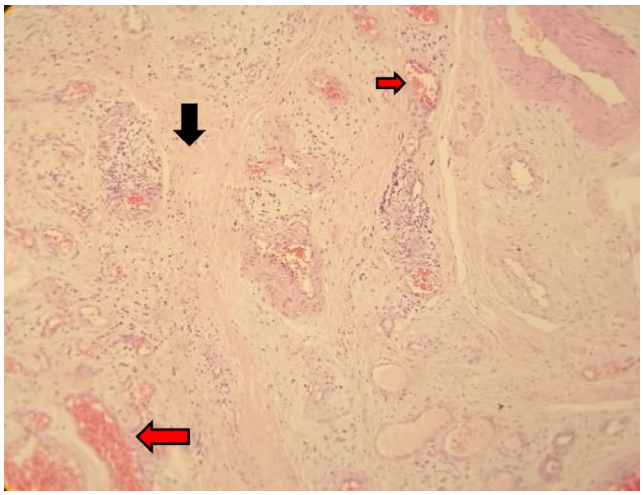

FIGURE 2 | Spindle stromal cells (black arrow) and vessels (red arrows) ranging from medium to large size.

On follow-up, the proptosis reduced to 10 mm as measured by Luedde's exophthalmometer (Figure 3). The patient was placed on a tapering dose of oral corticosteroids and advised to return for a follow-up after 3 months. The patient's condition improved with no recurrence of symptoms or complications, and the surgical outcome was favorable. The successful treatment, including careful surgical removal and appropriate postoperative management, led to a positive clinical outcome.

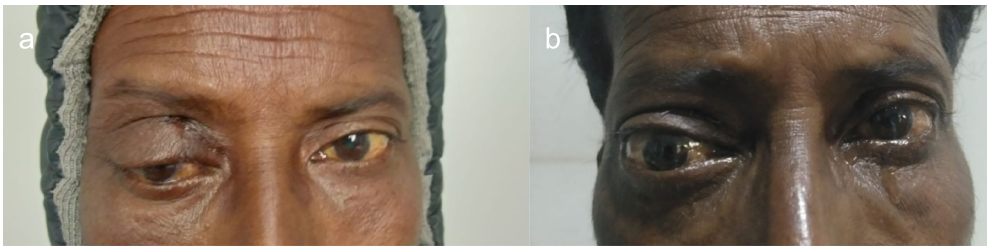

FIGURE 3 | Pre-surgical (a) and post-surgical (b) condition of the patient.

5 | Discussion

Orbital angiomyxoma, an extremely rare entity involving the orbit and periorbital cutaneous tissue, is a clinically significant differential for an orbital mass pertaining to the fact of its locally aggressive nature and risk of recurrence [2]. The patient in our study complained of pain occasionally, which was insidious, associated with forward bulging of the right eye without a decrease in visual acuity, but angiomyxoma of the orbit commonly presents as a painless palpable mass with or without decreased vision [4, 5]. So, the range of symptoms can be justified by the location of the orbital mass and the structures involved with it. It is usually seen in the adult and elderly age groups, with a mean age of 36.64, as with our case; however, cases have been reported in pediatric patients [6].

CT and MRI are a helpful diagnostic tools for planning the management of the case. CT scan findings commonly reported in other cases were heterogeneous masses with irregular borders and infiltration to adjacent structures like the medial orbital wall, ethmoidal sinus, medial rectus muscle, levator palpebrae superioris, or superior rectus [7]. Middle cranial fossa and cavernous sinus invasion of locally aggressive angiomyxoma has been reported by Hidayat et al. in two children [6]. This indicates a more aggressive nature of the disease in children.

Surgical excision is reported as the treatment of choice for the identified mass, while chemotherapy and radiotherapy have limited roles. For superficial angiomyxoma, local surgical excision can be done, while transcranial and orbitotomy approaches have been used for deep angiomyxoma [8]. Superior orbitotomy was done in our case, as the mass was located extraconal, where the entire encapsulated mass was excised. Due to the location and extension of the mass, complete excision is a challenging task requiring complicated approaches.

Histopathological examination is required for the establishment of the diagnosis and to rule out other differentials. Even though metastasis hasn't been reported in cases of it, the chances of recurrence and its affecting the vision of the patient require prompt and adequate management of the mass [4, 6].

6 | Conclusion

Orbital angiomyxoma is a rare but significant cause of orbital mass, presenting with proptosis and ocular symptoms. Timely diagnosis through imaging and histopathology, followed by surgical intervention, ensures favorable outcomes. Postoperative care and follow-up are crucial for preventing recurrence and ensuring recovery.

Author Contributions

Rajat Agarwal: conceptualization, writing – original draft. **Sajjad Ahmed Khan:** conceptualization, writing – original draft, writing – review and editing. **Priyanka Mishra:** writing – original draft. **Badri Prasad Badhu:** writing – original draft. **Prerna Arjyal Kafle:** writing – original draft. **Santosh Upadhya Kafle:** writing – original draft. **K. C. Ramesh Bikram:** writing – original draft.

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Consent

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

Data will be provided by the corresponding author upon reasonable request.

References

1. B. J. Sutton and J. Laudadio, "Aggressive Angiomyxoma," *Archives of Pathology & Laboratory Medicine* 136, no. 2 (2012): 217–221, https://doi.org/10.5858/arpa.2011-0056-RS.

2. S. Agrawal, A. Deora, S. Modaboyina, D. Das, M. S. Bajaj, and S. Sen, "Angiomyxoma of the Lower Eyelid With Orbital Extension: A Rare Entity and Review of Literature," *Annals of Pathology and Laboratory Medicine* 8, no. 4 (2021): C64–C70.

3. K. Haldar, I. E. Martinek, and S. Kehoe, "Aggressive Angiomyxoma: A Case Series and Literature Review," *European Journal of Surgical Oncology: The Journal of the European Society of Surgical Oncology and the British Association of Surgical Oncology* 36, no. 4 (2010): 335–339.

4. M. S. Bajaj, M. Mehta, and S. Kashyap, "Clinical and Pathologic Profile of Angiomyxomas of the Orbit," *Ophthalmic Plastic and Reconstructive Surgery* 27, no. 2 (2011): 76–80.

5. A. Mishulin, J. F. Lever, W. Porter, J. J. Servat, G. Gladstone, and E. Black, "Aggressive Glabellar Angiomyxoma With Orbital Extension,"

Orbit 31, no. 5 (2012): 361, https://doi.org/10.3109/01676830.2012.710925.

6. A. A. Hidayat, A. Flint, L. Marentette, et al., "Myxomas and Angiomyxomas of the Orbit: A Clinicopathologic Study of 6 Cases," *Ophthalmology* 114, no. 5 (2007): 1012–1019, https://doi.org/10.1016/j.ophtha.2006.12.022.

7. A. Pujari, M. S. Bajaj, S. Sen, and S. Goel, "Conjunctival Angiomyxoma-Rare but Needs Observation," *Canadian Journal of Ophthalmology. Journal Canadien D'ophtalmologie* 53, no. 2 (2018): e60–e62, https://doi.org/10.1016/j.jcjo.2017.07.005.

8. H. K. Yuen, W. Cheuk, F. O. Luk, C. S. Wat, K. C. Auyeung, and D. S. Lam, "Solitary Superficial Angiomyxoma in the Eyelid," *American Journal of Ophthalmology* 139, no. 6 (2005): 1141–1142, https://doi.org/10.1016/j.ajo.2004.12.019.