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Orbital primary intraosseous hemangioma in a three-month old infant: A case report

Babak Masoomian [a,*,1], Hassan Asadigandomani [a,1], Seyed Mohsen Rafizadeh [a], Shahriar Ghodous [b], Zohreh Nozarian [a]

[a] Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran
[b] Department of Medical Sciences, Guilan University of Medical Sciences, Rasht, Iran

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ABSTRACT

Purpose: To report a 3-month-old boy with rapid progressive orbital intraosseous hemangioma which was treated with excisional biopsy and orbital rim reconstruction.
Observation: A 3-month-old boy was referred with the aggressive growth of a mass on the right orbital region. The mass was noted to develop over 4 weeks. On presentation this firm nontender orbital mass measuring 5 × 5cm mimicked a more ominous malignancy. The spiral computed tomography scan showed a destructive mass with protrusion superiorly and inferiorly toward the orbital cavity and anterior cranial fossa. The patient underwent gross tumor resection and reconstruction of the orbital rim. Histology findings revealed an intraosseous hemangioma. There was no evidence of recurrence after 1-year follow-up.
Conclusion and Importance: Due to rapid progression, the patient's age, and lesion size, this case is unique. There were additional challenges regarding complete resection, intra-cranial extension, and significant blood loss in an infant. Therefore, in the face of rapidly progressing orbital tumors in infants, despite the very low prevalence of intraosseous hemangioma, this diagnosis should be considered.

1. Introduction

Intraosseous capillary hemangioma (IOH) is a rare benign vascular tumor presenting in the fourth and fifth decades of life.[1] It presents less than 1 % of all osseous tumors, and more than 50 % arise from the spine or skull.[1,2] Involvement in orbit is extremely rare,[1] and few cases have been reported. Most written articles belong to adult patients, so finding tumors in children and infants is extremely rare. Here, the authors present a 3-month-old infant with aggressive growth of an IOH treated successfully with surgery.

2. Case report

A 3-month-old boy presented with a history of a developing mass on the right superior orbital rim with aggressive growth over the previous 3 weeks. Examination revealed a hard reddish immobile mass on the right superior orbital margin that measured 5 × 5 cm in the greatest dimension. Complete ptosis was noted in the right eye secondary to the large mass (Fig. 1), which in turn prohibited examination of the anterior or posterior segments of the eye. There was no bruit or pulsation over this area.

The spiral orbital CT scan showed a lesion on the superior and supratemporal margin of the right eye protruding superiorly and inferiorly toward the orbital cavity and anterior cranial fossa. The central portion of the lesion had homogenous density, and destruction of the orbital rim was visible. The underlying brain parenchyma did not appear involved (Fig. 2). Based on these findings, the primary differential diagnosis was Ewing's sarcoma, but orbital metastases were also considered. Further systemic workup by a pediatrician failed to demonstrate more lesions or any evidence of primary malignancy.

Given the patient's history and clinical course, decision was made to proceed with excisional biopsy. A purplish, rounded tumor was surgically exposed through an eyebrow incision and gentle exploration. It was noted to arise from the orbital roof laterally. The salient portion of

* Corresponding author. Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Qazvin Square, Qazvin Street, Tehran, 14176-13151, Iran.
E-mail addresses: dr.masoomian@yahoo.com (B. Masoomian), hasanasadigandomani0800@gmail.com (H. Asadigandomani), mohsen_raf1354@yahoo.com (S.M. Rafizadeh), shahriarghodous@gmail.com (S. Ghodous), drnozarian@gmail.com (Z. Nozarian).
[1] These authors contributed equally to this work.

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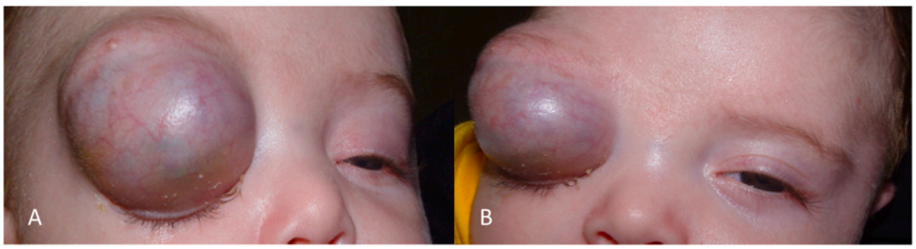

Fig. 1. Preoperative photograph showing a 5 × 5 cm firm and immobile lesion in right orbital region with complete ptosis.

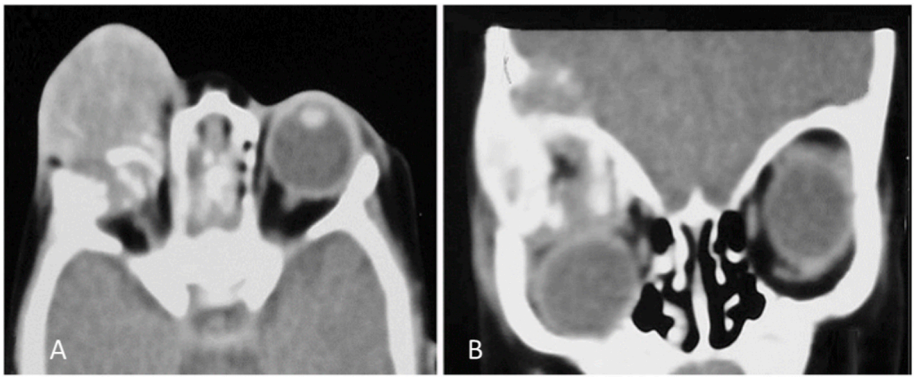

Fig. 2. (A) Spiral orbital CT scan (Axial view) showing a round huge non-homogeneous mass with bone particles in the center of the mass. **(B)** In (coronal view) inferomedial globe displacement with destruction of orbital roof is visible.

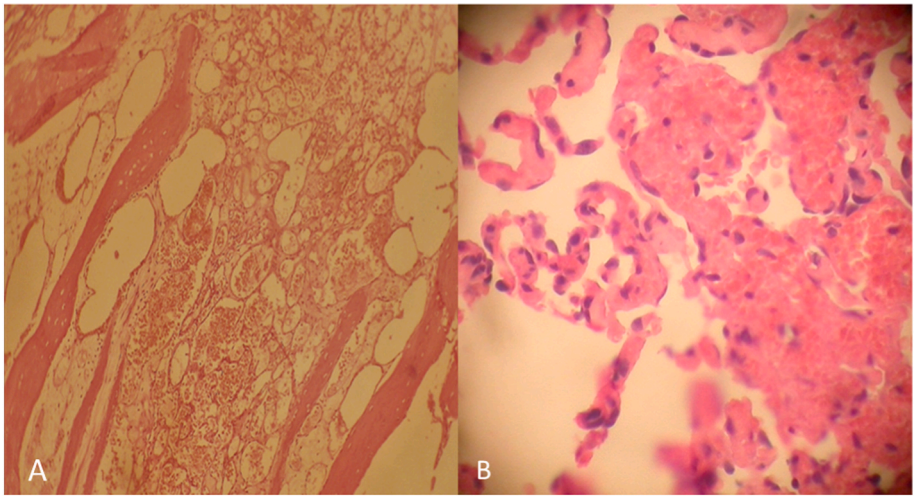

Fig. 3. (A) H&E stained show large, cystically dilated blood vessels filled with blood surrounded by trabecular bone (× 100). **(B)** H&E stained show papillary fronds lined by a single layer of endothelial cells proliferate within a vascular lumen. (× 400).

the mass was excised with an oscillating saw, incising the tumor at the level of the adjacent normal bone. Hemostasis was achieved using bone wax and compression. The tumor had destroyed the orbital roof laterally, and during surgery, the dura layer was exposed. Complete excision necessitated sacrificing portions of the lateral and superior orbital rims. The tumor was completely extradural, and a portion of the exposed dura was resected to ensure clear margins. The orbital defect was reconstructed with Medpor sheet implants without significant injury to the dura layer.

Grossly the specimen had yellowish color with a spongy feature. The specimen was sent in 10 % formalin solution after appropriate fixation and decalcified by immersing in an acid solution to prepare slices with reasonable thickness for processing and slide preparation.

Microscopic examination of Hematoxylin and Eosin (H&E) stained shows large, cystically dilated blood vessels surrounded by trabecular bone and lined by a single layer of endothelial cells within a vascular lumen. Also, some blood vessels show organized thrombosis and papillary endothelial hyperplasia-like changes, consisting of papillary fronds lined by a single layer of endothelial cells proliferating within a vascular lumen. Findings consistent with IOH (Fig. 3).

Twelve months after surgery, no signs of tumor recurrence were seen, and the patient remained symptom free. The patient was referred to the strabismus clinic due to right eye hypotropia, esotropia, and possible amblyopia (Fig. 4).

3. Discussion

In children, bone destruction can happen with several malignant

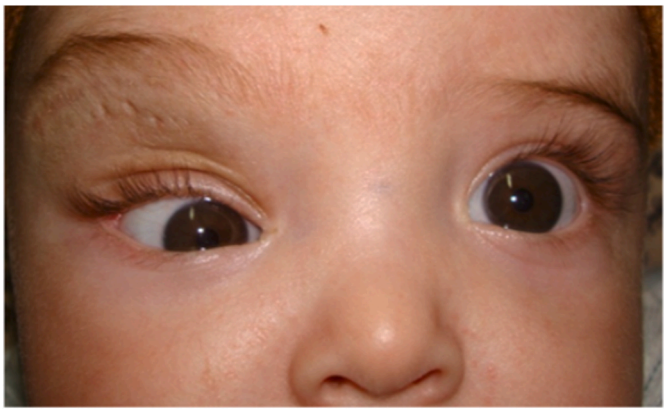

Fig. 4. Postoperative photograph showing hypotropia and also esotropia.

orbital lesions such as Ewing's sarcoma, Langerhans cell histiocytosis, metastasis (osteosarcoma, neuroblastoma), or rhabdomyosarcoma.[2–4] IOH and other vascular tumors are infrequent and, in total, comprise less than 1 % of all bone tumors in which almost 50 % of lesions arise from vertebra or skull.[3] In the skull area, the most affected bone is the parietal and then the frontal bone.[3] Based on published reports for IOH in general, there is a female preponderance on the order of 3:1 [3,4]. The primary age for the onset of the disease is in the 5th and 6th decades, but it has also been reported in young infants to very old ages.[4] In one published review article, from 49 patients with zygomatic IOH, only five patients were 18 years old or younger, and direct orbital involvement was reported in only seven cases.[5]

The tumor originates from vascular endothelial cells, but the pathophysiology of this tumor is still not completely clear. Although the history of trauma seems to play a role in its occurrence, most reports did not mention prior trauma.[6]

Clinically, these tumors present as a slowly growing, non-tender bony mass with subacute to chronic enlarging. Depending on the location and size of the lesion, diplopia, proptosis, globe displacement, and loss of vision may occur. Although there are different reports, the duration prior to presentation ranged from 1 month to 15 years, with a median of 12 months.[2,7] The differential diagnosis includes any firm, slow-growing mass of the skull or orbital bones with the normal overlying skin, such as fibrous dysplasia, aneurismal bone cyst, xanthoma of the bone, dermoid cyst, meningioma, osteoma, osteogenic sarcoma, and metastatic disease.[8] Imaging is essential, and radiographic features often suggest the diagnosis. CT scan with a bone window is the best method for finding bone lytic lesions in IOH. The classic descriptions for IOH are sunburst, honeycomb, and soap bubble. However, these findings are only sometimes present in patients with IOH and also may be seen in other osteolytic lesions.[9]

The histopathologic examination helps differentiate primary intraosseous hemangioma (PIOH: the presence of IOH in the absence of a known underlying disease) from other bone vascular malformations. According to the vessel's caliper, there are two subtypes of PIOH. Cavernous hemangioma (more common in the skull) contains large, thin-walled vessels lined by a single layer of endothelial cells. Capillary hemangioma (usually found in vertebrae bone) comprises finer and more vessels separated by normal bone tissue. These two types of PIOH usually have similar imaging findings and differentiation made only with histopathological evaluation. The capillary subtype of PIOH is less common (20 % vs. 80 %), and compared to the other one, the mean age at the onset of symptoms was also younger (28 years vs. 49 years). There is some difference in the behavior of the two subtypes. Capillary types have rapid growth behavior and are associated with a greater likelihood of recurrence. Conversely, surgery for cavernous types was more likely to be complicated by significant bleeding.[10] Immunohistochemical analysis for vimentin, CD31, and Factor VIII may be appropriate and useable in special cases.[2]

Patients with a benign course of the disease can be observed without debilitating symptoms, however surgery is reserved for patients with mass effects, bleeding, or severe aesthetic concerns. Total en-bloc excision with a small margin of normal tissue has been advocated as the treatment of choice. There is the risk of significant bleeding during surgery; some references supported preoperative angiography guided embolization.[11] Fortunately, we had no severe hemorrhage or considerable blood loss during the operation. The case we present has 12 months of follow up, however recurrence after complete resection in these tumors is uncommon.[1,12]

In summary, IOH is an extremely rare and benign vascular tumor commonly present in the fourth and fifth decades of life. It is a slow-growing mass, often on the orbital rim, and the lesion's location within the orbit will determine the tumor-related symptoms. Treatment is most commonly surgical excision, with or without preoperative embolization, to reduce intraoperative bleeding, thus surgical removal of these lesions requires additional preoperative considerations that should be discussed with the patient including the possibility of significant blood loss.

4. Conclusion

According to our best knowledge, the presented patient is one of the few infants with IOH in the orbital region, which differing from the normal presentation of a slow and gradual growth, our case highlights rapid growth and visual disability.

Patient consent

We obtained permission from the patient's parents in the consent form to publish her information without mentioning her name.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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