



CASE REPORT

Hemangioendothelioma of the orbit in a 3-month-old infant[☆]

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KEYWORDS

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Summary Hemangioendothelioma of bone, a locally aggressive vascular neoplasm, rarely affects the facial bones. Even less common is a presentation of this tumor affecting the orbit. Excision of orbital rim tumors and consequential reconstructions in infants are especially challenging because calvarian bone grafts are not available and because of the potential for orbital deformity and facial scarring. A case of a 3-month-old female with a rapidly enlarging tumor of the inferior orbital rim and maxilla is presented. Resection was accomplished with a unique modified subciliary skin incision; orbital rim reconstruction was accomplished with dermis allograft (Alloderm[®]) only.

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1. Introduction

Hemangioendothelioma of bone is a locally aggressive vascular neoplasm of intermediate clinical behavior, between benign hemangioma and angiosarcoma. Facial bones are rarely affected. There have been only two previous reports of hemangioendothelioma affecting the orbit. Excision of orbital

tumors in infants and orbital reconstruction is challenging. We report a case of a 3-month-old female with a rapidly enlarging hemangioendothelioma of the inferior orbital rim and maxilla. The tumor was excised via an extended subciliary incision; reconstruction was carried out with dermis allograft (Alloderm[®]) only with excellent cosmetic result.

2. Case report

A 3-month-old female presented with a rapidly enlarging mass of the left inferior orbital rim and cheek causing mild erythema of the overlying skin. Physical examination revealed a firm mass with smooth surface at the medial aspect of the inferior

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Fig. 1 Preoperative CT showing characteristic lytic, "cystic" lesion.

orbital rim and cheek with slight upward displacement of the globe.

Computed tomography (CT) showed a "cystic," lytic, non-enhancing, well circumscribed, focally infiltrative mass (**Fig. 1**). Magnetic resonance imaging (MRI) revealed a multilobulated, contrast-enhancing mass filling the left maxillary sinus,

extending to the inferior orbital rim, floor of the orbit, soft tissues of the cheek, and to at least one tooth bud. There was displacement of the inferior rectus and the globe.

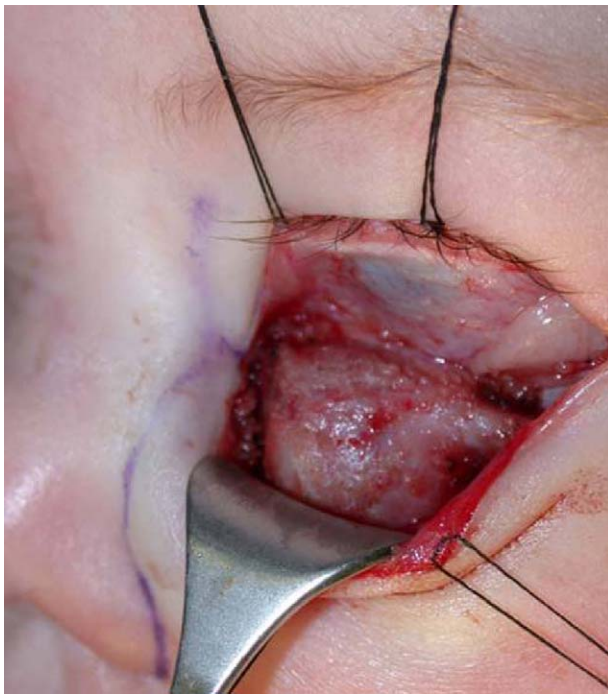


Fig. 2 Extended subciliary approach; tumor occupying the inferior orbital rim and maxillary sinus.

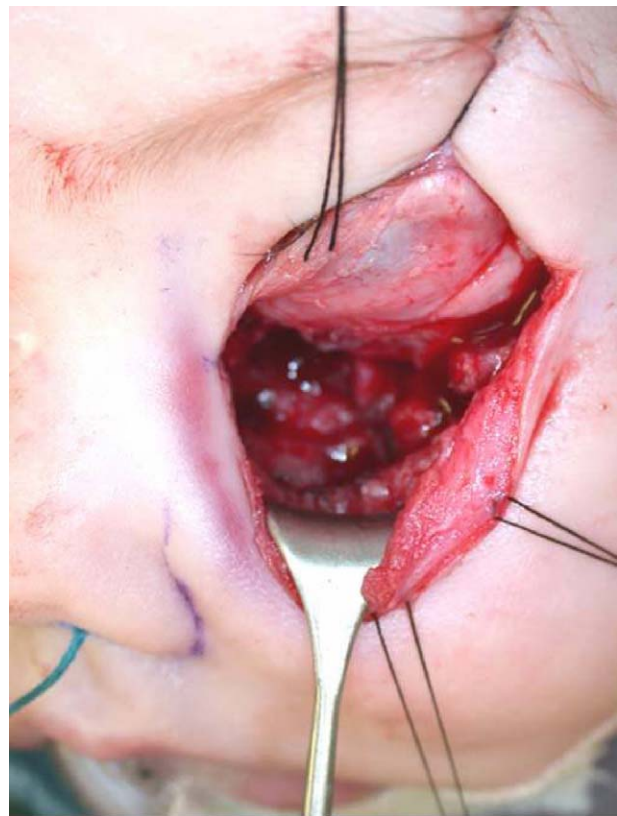


Fig. 3 Surgical defect.

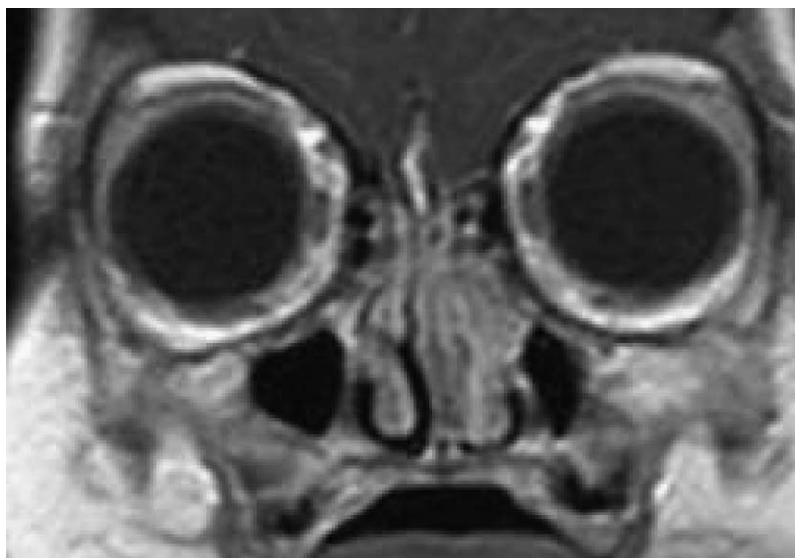


Fig. 4 MRI at 20 months follow-up with no evidence of recurrence.

Biopsy was performed via a sublabial incision with significant blood loss. Pathologic evaluation revealed hemangioendothelioma. Resection was performed through a subciliary incision. The incision was extended medially and inferiorly along a line of fusion between the nasal skin and the cheek skin. This provided a wide exposure of the medial aspect of the inferior orbital rim and maxillary sinus (Fig. 2). Complete tumor resection required resection of at least 50% of the medial aspect of the orbital rim with most of the orbital floor and medial wall of the maxillary sinus (Fig. 3). The orbital rim and floor were reconstructed with a thin sheath of Alloderm[®] without bone grafting.

Pathologic examination of the specimen revealed a highly cellular tumor, with closely packed vessels producing a solid cellular appearance. Immunohistochemical studies using CD31 and SMA demonstrate

individual vessels showing no significant endothelial multi-layering or atypia, consistent with Grade I hemangioendothelioma of bone. At 20 months follow-up, there was no radiological evidence of recurrence (Fig. 4). The orbital contour is well preserved, with no noticeable scar (Fig. 5).

3. Discussion

Most hemangioendotheliomas occur in long bones. These tumors are exceedingly rare in the head and neck, with only isolated case reports of involvement of the frontal, temporal and mandibular bone. There have been only two previous reports of hemangioendotheliomas involving the orbit. This tumor has no known age or sex predilection.

Histologically, hemangioendotheliomas are described as intermediate in appearance, between benign hemangiomas and conventional angiosarcomas. The tumor is comprised of groups of irregular vascular elements lined by immature endothelial cells with prominent anaplasia. All hemangioendotheliomas are positive for at least one endothelial marker (CD31, CD34, factor VIII, Ulex europaeus). Unni et al. in 1971 proposed a grading system for hemangioendotheliomas of bone (I–III). This grading is based on the degree of vasoformation, nuclear anaplasia and number of mitotic figures. Grade I lesions have abundant vascular spaces lined by cells with mild atypia. Tumor grade is probably a predictor of biological behavior [1].

Hemangioendotheliomas affect all age groups. Multifocality is present in 9–14% of cases. In the orbit, hemangioendothelioma presents as a very rapidly enlarging mass (in this case the tumor



Fig. 5 Twenty months follow-up. Orbital contour is well preserved with no noticeable scar.

doubled in size within 2 weeks) with edema or erythema of the overlying skin. The tumor is highly vascular, with significant bleeding encountered on biopsy. Unlike the behavior of rhabdomyosarcomas, hemangioendotheliomas have a pushing, or mass, effect on neighboring structures, rather than invasive behavior. On imaging, these tumors are found to be lytic, multiloculated, expansile lesion with localized invasion into adjacent tissues.

Vascular lesions are the second most common orbital tumors in childhood, comprising approximately 15% of cases. The vast majority of these tumors is benign and includes cavernous hemangioma, orbital varix, lymphangioma, capillary hemangioma, hemangiopericytoma, and arteriovenous malformation. Malignant vascular lesions in this location are rare. A 2002 review of the literature by Triantafyllidou yielded only six reported cases of angiosarcoma originating from the maxilla and two cases from the orbit/maxillary sinus. All of these lesions were reported in the third decade of life or later [2].

Complete surgical resection is the primary treatment modality for hemangioendotheliomas of bone; adjuvant treatment is dependent on histological grading. For grade II, radiation is used. Grade III tumors are treated as sarcomas, with radiation and chemotherapy. Grade I tumors do not require adjuvant treatment. Local recurrence is a significant problem with hemangioendotheliomas of higher malignant potential necessitating regular follow-up with MRI [3].

Removal of more than half of the inferior orbital rim, if not reconstructed, may cause noticeable orbital deformity. Reconstruction is often accomplished with split thickness calvarian bone graft. In a 3-month-old infant, bone graft is not readily available. In order to completely excise the tumor, a wide exposure of the medial aspect of the inferior orbital rim and maxilla was achieved by extending the subciliary incision along the skin fusion line between the lateral nasal and cheek regions. A skin flap was elevated separately from the orbicularis

oculi muscle. The skin was retracted and the muscle was elevated off the tumor and the orbital rim. In the only previous report of a similar case, a lateral rhinotomy incision with consequential facial scarring was utilized [4]. We have reconstructed the orbital rim and floor with dermis allograft only. At 20 months follow-up, the facial and orbital contour is well preserved, with no noticeable scar. Bone reconstruction of the orbital rim may be needed in the future if a noticeable deformity will develop as the facial bones further develop, and the overlying soft tissues thin down.

4. Conclusion

Hemangioendothelioma of the orbital bone is a rare vascular tumor with only two previously reported cases. These tumors have an intermediate clinical behavior, between hemangiomas and angiosarcomas. The case presented is noteworthy because of the complexity associated with surgical management of orbital tumors in infants and the unique surgical reconstruction used. Extensive orbital rim and floor resection defects were reconstructed with dermis allograft only, resulting in excellent facial and orbital contour at 20 months follow-up.

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