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Endovascular Preoperative Embolization of Orbital Hemangiopericytoma With n-Butyl Cyanoacrylate Glue

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Abstract

Hemangiopericytoma is an uncommon neoplasm that may present in myriad locations, including the lower extremities, pelvic area, and the head and neck area, including the orbit.¹ Orbital hemangiopericytoma is often described as synonymous with orbital solitary fibrous tumor, giant cell angiofibroma, and fibrous histiocytoma, as they all belong to a spectrum of collagen-rich fibroblastic tumors that are often CD34-positive and have overlapping histopathologic features.² Many cases of orbital hemangiopericytoma have been reported in the literature along with various surgical approaches, long-term outcomes, and techniques to manage recurrence; however, few have discussed preoperative embolization.^{1,3–5} Intraoperative hemorrhage is a concern in both the congenital and the adult form of these cases^{6,7} and may be an indication for preoperative embolization. A unique case of preoperative embolization was presented with n-butyl cyanoacrylate for surgical resection of a large orbital hemangiopericytoma in a 58-year-old woman.

CASE REPORT

In accordance with HIPAA policy, the identity of this patient will remain anonymous, and explicit written permission was given to the authors to publish the photographs of her face.

A 58-year-old Caucasian woman from a rural area presented to an outside clinic complaining of OS proptosis. Her symptoms started 2 years earlier with discomfort and injection of the OS. Over the most recent 6 months, she began noting progressive exophthalmos and a reddish mass nasal to her OD. She complained that her central and peripheral vision had decreased. The patient reported orbital pressure and anxiety about her appearance (Fig. 1A). She denied any other past medical problems or current associated neurological problems, including headaches, seizures, imbalance, or weakness.

The patient was referred to the institution for evaluation and surgical management of this orbital mass. With regard to the OS, she was found to have a Snellen visual acuity of

20/200, a relative afferent pupillary defect, exophthalmos (Hertel measurement 19 mm on the right side, 31 mm on the left with base 118 mm), temporal displacement of the globe, lagophthalmos of 5 mm, severe restriction of extraocular movements, a normal intraocular pressure, and pallor of the optic nerve head without acute papillitis. The cornea appeared healthy with no uptake of fluorescein dye. Goldman visual field examination showed an enlarged blind spot and decreased sensitivity to the I-4 isopter.

MRI/magnetic resonance angiography (MRA) of brain and orbits were performed; MRA failed to show any signs of intracranial or intraorbital fistula, thrombosis, or aneurysm and thus provided no evidence for venous congestion causing this mass. MRI showed a large, 4.8-cm extraconal mass in the left orbit, causing significant lateral displacement of the optic nerve and globe and proptosis of the globe (Fig. 2A,B). There were multiple signal voids suggestive of large feeder vessels noted on the T2-weighted MRI (Fig. 2A,B). The mass was heterogeneously enhancing with contrast and in the delayed images, there was continued washout of contrast from the lesion (Fig. 2C,D). These radiographic findings were thought to be consistent with a highly vascular orbital hemangioma or hemangiopericytoma.

After consultation with the Endovascular Neurosurgery service, and discussion with the patient, the decision was made for the patient to undergo cerebral angiography and to attempt embolization of the lesion preoperatively. A diagnostic cerebral angiogram demonstrated a hypervascular orbital tumor with multiple feeder vessels arising from the third segment of the left ophthalmic artery as well (Fig. 3A) as from the nasal–frontal branch of the internal maxillary artery. Based on the extensive amount of vascularity, the patient underwent preoperative endovascular embolization. With a (1.2-French nonolive tip) Magic microcatheter (Bolt Inc., Montgomery, France) selectively placed 5 mm distal to the origin of the central retinal artery, the orbital tumor was embolized using diluted n-butyl cyanoacrylate glue (n-BCA; Cordis-Codman, Raynham, MA) in ethanediol oil (15% concentration; Fig. 3B). In an attempt to facilitate distal migration of the n-BCA and avoid the reflux of the embolic agent in the central retinal artery, 5% dextrose water was infused concomitantly in the proximal guide catheter. At the end of the embolization, a complete devascularization of the tumor had been achieved with no compromise of the central retinal artery (Fig. 3C). She was monitored in the neurosurgical intensive care unit overnight while receiving 250 mg of intravenous methylprednisolone every 6 hours and was taken to the operating room the next morning. Immediately after embolization, the patient subjectively reported improvement in her orbital pressure and in her visual acuity.

Intraoperatively, the lesion appeared reddish blue, hypervascular, fibrotic, and encapsulated. An anterior “eyelid-split” orbitotomy was performed to completely resect the encapsulated mass, and care was taken to avoid damage to the superior oblique and medial rectus muscles, and to the superior orbital neurovascular bundle. Estimated blood loss was 20 ml intraoperatively. The patient was discharged home the next day in good condition.

Pathologic analysis of the lesion demonstrated a well-circumscribed mass composed of spindle cells in loosely arranged fascicles with extracellular collagen deposits and telangiectatic vessels. There were spaces associated with coagulative necrosis and myxoid degeneration, consistent with the effects of embolization, per the pathology service (Figs. 4 and 5).

Immunohistochemical staining for CD34 was grossly positive. With these morphologic and immunohistochemical findings, the diagnosis of hemangiopericytoma was made.

Two months postoperatively, the patient is doing well with no signs of recurrence (Fig. 1B). She experiences mild residual diplopia with significant improvement in her visual acuity to the 20/50 level.

DISCUSSION

Hemangiopericytoma, or more broadly, solitary fibrous tumor, is an uncommon spindle cell neoplasm of the orbit. Hemangiopericytoma makes up 1% to 3% of all biopsied orbital lesions.⁸ Due to the paucity of literature about the behavior of these orbital tumors, the clinical course and prognosis of these patients is not well understood. As can be seen in this case, they can clinically and radiologically resemble other common tumors of the orbit such as lymphangiomas and hemangiomas. This can be challenging when choosing therapy (or choosing to monitor) for these patients. According to Eivazi et al.,⁹ the distinction must be made between vascular malformations and hemangiomas. Furthermore, the hemodynamic character must be understood, to anticipate the risk of hemorrhage and the possible need for preoperative embolization. Distensibility of the lesion, the extent and type of flow can be best determined by integration of clinical clues, such as the Valsalva maneuver, and imaging modalities (as exemplified in this case).¹⁰

Total resection is the definitive treatment to reduce likelihood of recurrence, malignant transformation, or need for reoperation in cases of hemangiopericytoma.^{11,12} Other reports have described the use of preoperative radiation,⁸ adjunctive proton beam therapy,¹³ and adjunctive brachytherapy for primary and/or recurrent hemangiopericytomas.¹⁴ As intraoperative hemorrhage can be a risk in these cases,^{6,7} some reports have discussed the use of preoperative embolization. Methods for embolization have included polyvinyl alcohol, Onyx (eV3-Covidian, Irvine, CA), and particulate embospheres (150–300 μm).^{1,4,5} The use of n-BCA for preoperative embolization of orbital hemangiopericytoma was reported. The use of n-BCA to embolize vascular lesions in the orbit has been reported in the past. Couch et al.¹⁵ discussed 4 cases of orbital varices that were treated with preoperative percutaneous injection of n-BCA to embolize the varicosities prior to resection. Tsai¹⁶ described a case of an orbital varix that was first exposed surgically and then was embolized with n-BCA preceding resection. Lacey et al.¹⁷ reported intraoperative venography and n-BCA embolization of orbital venous malformations prior to surgical resection. The variation described in these latter studies is that they surgically exposed the venous malformation before injection, whereas the authors opted to embolize 1 day prior to surgery. The efficacy of this method was seen surgically, as this very large and vascular mass was able to undergo total resection with minimal blood loss. Onyx and n-BCA are both viable options for embolization; however, there are differences between the agents. It is very important to form a “plug” with Onyx reflux at the tip of the microcatheter before distal migration of the Onyx occurs.¹ This technique may have been risky in this case because of the short distance between the origin of the central retinal artery and the tip of the microcatheter. There is also a risk with the use of n-BCA associated with possible early precipitation of the glue. This risk was ameliorated with the technique used (see below); however, further study will be

needed to compare these 2 agents. Jeeva et al.⁵ described the successful use of embosphere particles, which also offer good control and low risk of reflux. However, Wang et al.¹⁸ recently reported the increased risk of hemorrhage in using embospheres when compared with n-BCA in hypervascular pediatric brain tumors.

Diluted n-BCA with continuous 5% dextrose water infusion in the guide catheter was used to delay the time to precipitation. Moore et al.¹⁹ used n-BCA to treat dural arteriovenous fistulas where distal penetration of the glue was desired and the microcatheter was placed far from the fistula site. Diluted glue is thought to delay the precipitation time because the diluted glue delays the contact of n-BCA with ionic solution (blood). The use of the 5% dextrose solution in the proximal guide catheter is thought to delay the precipitation of the glue by diluting the incoming ionic blood with nonionic dextrose solution. In this patient, the authors were able to fill the trifurcation of the ophthalmic artery with no proximal glue reflux. The glue was allowed to penetrate deeply into small feeder vessels within the tumor. The final angiogram does confirm the patency of the central retinal artery with a patent small stump of the ophthalmic artery distal to it. The risk of embolization includes inadvertent embolization of the central retinal artery. This risk should always be weighed against the benefit of minimizing intraoperative blood loss.

Thus, an extraordinary case of a very advanced hemangiopericytoma was report, which was totally resected with minimal blood loss after preoperative embolization with n-BCA glue, using the proximal dextrose push technique.

CONCLUSIONS

Hemangiopericytoma, or solitary fibrous tumor, is an uncommon orbital neoplasm that can present in a very advanced manner with the risk of hemorrhage which can be managed by preoperative embolization with n-BCA glue followed by total resection.

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FIG. 1.

A, External photograph of patient exhibiting marked proptosis and large intraorbital mass. The eye appears to be displaced laterally. The conjunctiva and the eyelid are bluish in color reflecting increase vascularity. **B**, Patient in the second postoperative month showing dramatic improvement in proptosis, mass effect, exposure, and cosmesis with no signs of recurrence.

FIG. 2.
A and B, Coronal T2 MRI sequence of the orbits showing a 4.8-cm intraconal lesion with pressure effect and lateral displacement of the left eye. **C and D,** Axial T1 MRI sequence postcontrast of the same lesion.

FIG. 3.

A, Selective catheterization and angiography of the ophthalmic artery with a 1.2-French microcatheter. The tumor appears to be highly vascular with multiple large feeder vessels raising from the ophthalmic artery trifurcation. The central retinal artery origin (*large arrow*) appears to be originating 5 mm from the ophthalmic artery trifurcation (*small arrow*). n-Butyl cyanoacrylate (n-BCA) was injected from the ophthalmic artery trifurcation (*small arrow*). **B**, Saved road-map image at the end of the glue injection through the ophthalmic artery, the glue cast filling the distal vessels within the orbital tumor. There is no n-BCA glue reflux proximal to the ophthalmic artery trifurcation. **C**, Internal carotid artery angiogram postembolization of the orbital tumor. The tumor appears to be completely devascularized, the central retinal artery is patent, and the normal choroidal blush of the globe is seen.

FIG. 4.
Gross anatomical image of the well-encapsulated orbital tumor status post resection.

FIG. 5.
A and B, Hemangiopericytoma. Low and high magnification sections (hematoxylin-eosin) from excised orbital mass. There is a highly cellular lesion, with spindle cells and telangiectatic vessels. Embolic agent (*arrow*) seen within small tumor vessels.