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Intraosseous Hemangioma of the Frontal Bone. Report of a Case and Review of the Literature

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Intraosseous hemangiomas of the maxillofacial region are rare lesions that constitute less than 1% of all osseous tumors. A review of the literature on intraosseous hemangiomas of the skull and facial bones showed a limited number of publications, much of which were largely limited to case reports.

This case report summarizes the workup and surgical treatment of a 39-year-old woman with an intraosseous hemangioma of the left frontal bone. The histology, treatment, and literature are reviewed.

Report of Case

The patient was a 39-year-old woman who was referred to the Department of Oral and Maxillofacial Surgery at the Harborview Medical Center (HMC; Seattle, WA) by her general dentist for evaluation of left facial pain and left forehead swelling of 3 months' duration. She described the pain as dull, with intermittent episodes of shooting pain, and a sensation of constant pressure over her left forehead. She denied any history of trauma to the area or any changes to her vision. The patient's medical history was notable for asthma, depression, and cytochrome P450 mutation. She denied any asthma attacks in the past or the need for an inhaler, and her depression was well controlled with escitalopram. She had a negative surgical history and denied having any family members with similar vascular lesions. She smoked 1 pack of cigarettes per day while in her 20s but quit shortly thereafter, and she denied any history of other drug or alcohol use.

Physical examination showed a 2- × 2-cm fixed bony mass located on the left forehead, approximately 1.5 cm superior to the left eyebrow (Fig 1). Her neurologic examination and further examination of her head and neck were negative for pathology. Maxillofacial computed tomography (CT) showed a mixed radiolucent or radiopaque lesion of the left frontal bone with expansion through the inner and outer cortices and expansion into the superior orbit (Fig 2). Differential diagnosis was primarily osteosarcoma versus hemangioma and magnetic resonance imaging (MRI) was recommended. MRI showed a heterogeneous T2 hyperintense and mildly T1 hyperintense lesion in the left frontal diploic space with mild scalloping of the adjacent cortex measuring 2 cm transversely × 1.3 cm anteroposteriorly × 2.2 cm in the superoinferior direction (Fig 3). There was expansion of the outer table with a mild deformity of the supraorbital aspect of the left frontal bone as a result of the lesion and avid enhancement of the lesion after administration of contrast. These findings were noted to be more consistent with an intraosseous hemangioma than with an osteosarcoma. The patient was preoperatively evaluated by neurosurgery and oral and maxillofacial surgery and surgical options were discussed in detail. She was given the option of complete resection with immediate reconstruction (understanding that the diagnosis was not confirmed) versus complete resection with reconstruction at a later date once the diagnosis was confirmed. The patient opted for complete resection with immediate surgical reconstruction.

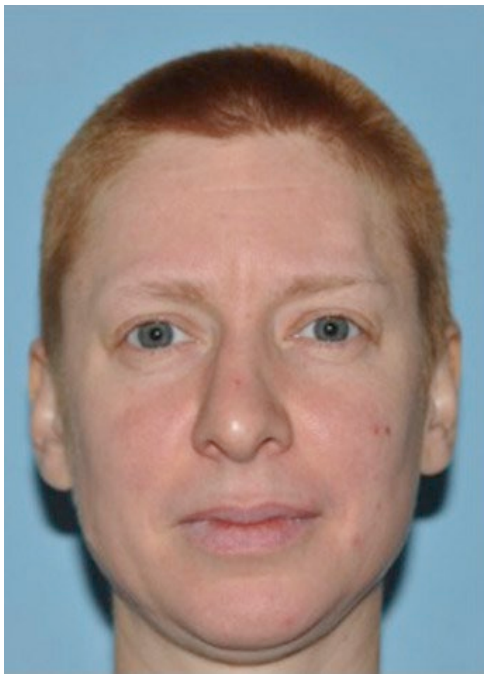

Figure 1. Preoperative clinical photograph showing left frontal intraosseous hemangioma.

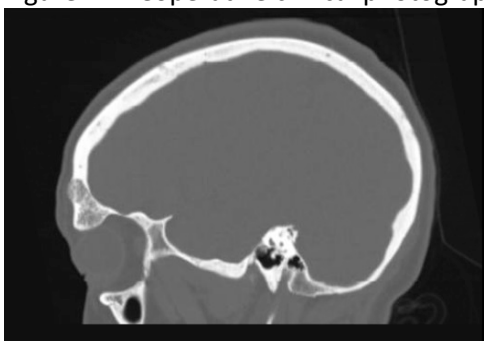

Figure 2. Sagittal computed tomogram showing intraosseous hemangioma.

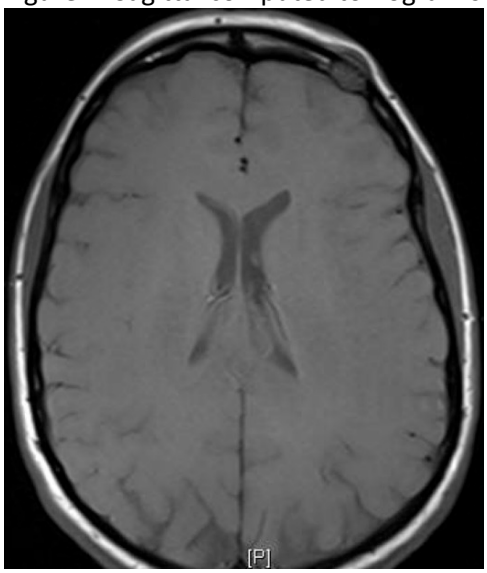

Figure 3. Axial magnetic resonance image showing intraosseous hemangioma.

CT was used to conduct virtual surgical planning to create a porous polyethylene (MEDPOR) customized implant (Stryker, Kalamazoo, MI). Given the challenges of creating a custom preoperative

implant of a defect that had not yet been created, additional volume was incorporated into the implant to allow for intraoperative adjustments. Of note, an intraoperative 3-dimensional printed implant of the surgical defect would have been ideal but is not currently available at the HMC. The porous polyethylene implant was designed according to resection of the lesion, with the medial defect margin approximately 20 mm from the frontonasal suture and the lateral defect margin 21 mm from the coronal suture. The implant was fabricated using a porous polyethylene polymer at a thickness of 4.5 mm (Fig 4).

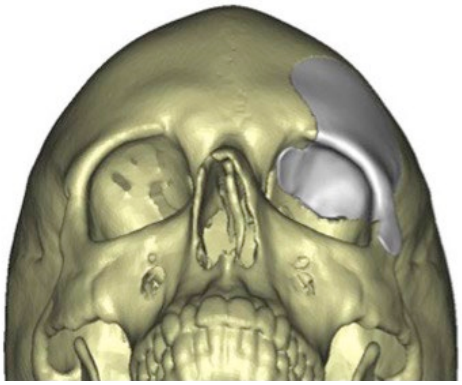

Figure 4. Three-dimensional reconstruction after MEDPOR implant placement.

The patient was scheduled for angiography followed by resection of the lesion with coronal flap, left frontal craniotomy, orbital ostectomy, and resection of the intraosseous hemangioma from the skull and frontal bone, with the assistance of intraoperative stereotactic navigation. A complete diagnostic cerebral angiogram showed that the lesion was not particularly hypervascular but was supplied by branches from the left middle meningeal artery. No other vascular abnormalities were noted and there was no indication for embolization. She was taken to the operating room the following day with neurosurgical assistance for removal of the lesion with subsequent reconstruction. A coronal flap was raised by the neurosurgery team and the intraosseous hemangioma was visualized (Fig 5). Using the stereotactic navigation system, the border of the mass was marked and the left frontal craniotomy and orbital osteotomy were performed for en bloc resection of the bony lesion. Once clear margins had been delineated under image guidance, a bur hole was fashioned in the frontal bone just above the temporal line and posterior to the orbital floor to expose the frontal dura. Then, a craniotome was inserted and used to cut the frontal margins surrounding the hemangioma and a reciprocating saw was used to make the cuts through the orbital roof. A mallet and an osteotome were used to separate any residual bony attachments and the specimen was delivered en bloc with the frontal dura remaining intact (Fig 6).

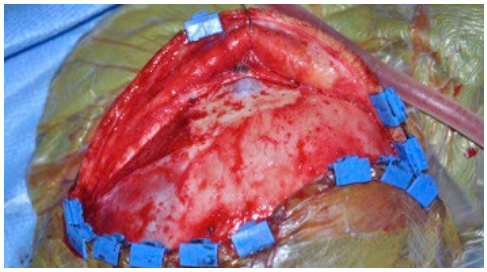

Figure 5. Intraoperative bird's-eye view of intraosseous hemangioma.

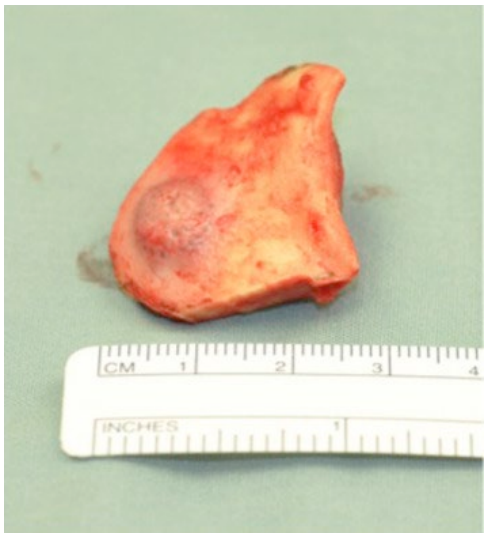

Figure 6. Surgical specimen after en bloc resection.

Attention was turned to the defect, where a contouring bur was used to shape bony margins. Multiple fine adjustments were made to the implant and to ensure a proper fit into the defect site. Once the implant was fitted and contours were noted to be satisfactory, the orbital roof aspect of the implant was thinned to prevent inadvertent pressure on the surrounding anatomy. After the fitting process was complete, the implant was cleaned and placed in bacitracin solution. Then, the implant was placed into position and secured with Stryker 1.2-mm square midface plates using 4-mm monocortical screws (Fig 7). The case was turned back to neurosurgery for reapproximation of the pericranium and closure of the coronal flap.

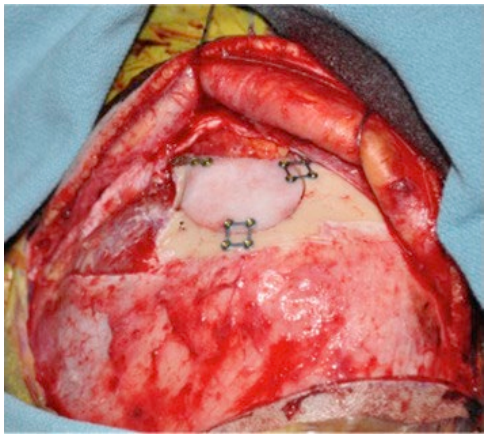
Figure 7. Intraoperative bird's-eye view of surgical site after reconstruction.

The patient was admitted postoperatively and was given Decadron (dexamethasone; Merck, Kenilworth, NJ) 8 mg every 8 hours for 48 hours. She had an uneventful hospital stay and was discharged on the third postoperative day. She was examined in the outpatient clinic 3 weeks later and was noted to have mild residual edema. She did not have any neurosensory deficits or changes to her vision from baseline. She was examined again after a 6-week postoperative period. At that time, there was complete resolution of the edema and optimal healing of the surgical site. Repeat CT displayed postsurgical changes but no evidence of recurrence or abscess. The porous polyethylene implant

successfully re-created the natural contours of the area of resection and left an esthetically pleasing result (Fig 8).

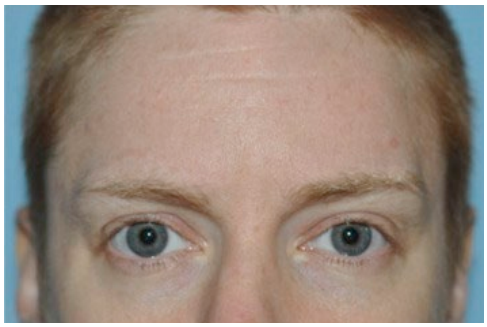

Figure 8. Postoperative clinical photograph at 6 weeks.

Macroscopic and Histopathologic Examination

The resected bone specimen measured approximately 3.5 × 2.5 × 2 cm. Quadrisected sections of the bone specimen showed dark red hemorrhagic spongy tissue bound by dense cortical bone (Fig 9). Decalcified sections stained with hematoxylin and eosin showed an intraosseous lesion composed of numerous thin-walled cavernous vascular channels separated by bony trabeculation (Fig 10). The vascular channels were lined by a single layer of flat endothelial cells and were often engorged with erythrocytes (Fig 11). Verhoeff-Van Gieson stain failed to visualize an elastic lamina around these vascular channels, thereby implicating venous differentiation (Fig 12). Cytologic atypia, cell crowding, mitotic activity, and giant cells were not present. A diagnosis of an intraosseous cavernous hemangioma was given.

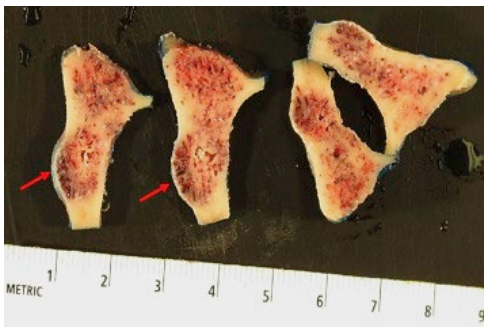

Figure 9. Quadrisected gross specimen showing a spongy red mass occupying the cancellous bone. Note the cortical expansion (arrows).

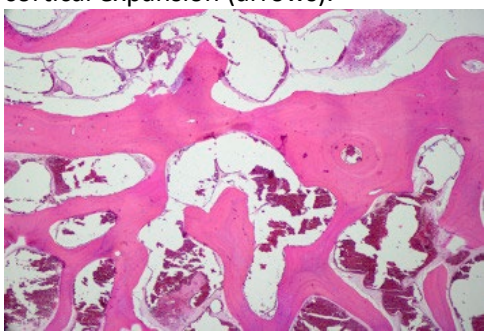

Figure 10. Cavernous vascular channels separated by trabeculae of cancellous bone (hematoxylin and eosin stain; magnification, ×4).

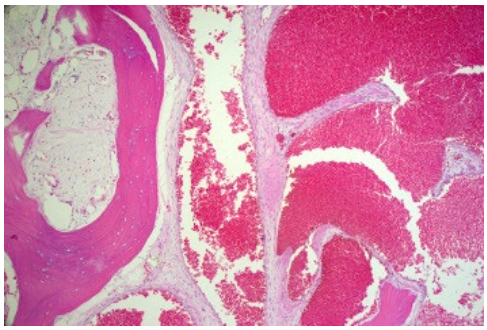

Figure 11. Vascular channels are lined by a single layer of flat endothelial cells and are engorged with erythrocytes (hematoxylin and eosin stain; magnification, ×10).

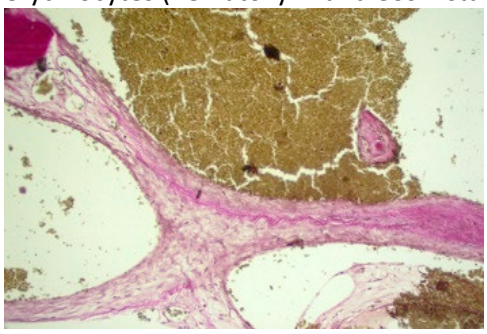

Figure 12. Cavernous vascular channels lack an elastic lamina (Verhoeff-Van Gieson stain; magnification, ×20).

Discussion

This case report summarizes the workup and surgical treatment of a 39-year-old woman with an intraosseous hemangioma of the left frontal bone. Intraosseous hemangiomas are rare lesions that constitute less than 1% of all osseous tumors and most commonly involve the frontal and parietal bones, nasal bones, zygoma, maxilla, and mandible.[1]

Vascular malformations represent true errors in the development of the embryonic vasculature.[2] They are developmental in nature and typically grow proportionally with the host. They often manifest arteriovenous shunting, vascular ectasia, and thrombosis. They are predominantly soft tissue lesions and bone involvement is secondary.[3] The 2014 classification of vascular anomalies by the International Society for the Study of Vascular Anomalies (ISSVA) deals largely with vascular tumors and vascular malformations of infancy and childhood. In addition, correlation with vascular bone lesions is not standardized.[4] A previous study on vascular lesions of bone in children, adolescents, and young adults recommended that the ISSVA classification be applied to vascular lesions of bone just as for those originating in the skin, soft tissue, and viscera. Although the study restricted the cases to bone lesions, it included only patients no older than 25 years. This apparently is in keeping with the developmental nature and growth of vascular malformations proportional to the growth of the host with closure of epiphyseal growth plates by this age.[5] Solitary vascular lesions of bone diagnosed as hemangiomas have a peak incidence from the fourth to sixth decades of life, with the flat bones of the calvarium accounting for nearly 50% of cases. Other common sites include the vertebral bodies and long tubular bones.[3] The present patient was 39 years of age. She presented with a solitary expansile lesion of the frontal bone with no history of childhood vascular anomalies or trauma to explain the findings. Clinical examination and diagnostic cerebral angiography did not show soft tissue involvement. A diagnosis of an intraosseous hemangioma was made by correlating the clinical presentation and cerebral

angiographic and histopathologic findings with those described in literature such as the 2013 World Health Organization classification of tumors of soft tissue and bone[6] (Fig 13).

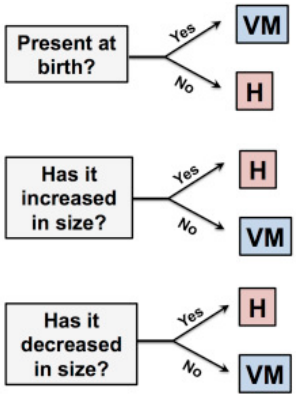

Figure 13. Hemangioma versus venous malformation. H, hemangioma; VM, venous malformation. From Waner M, Suen JY: Hemangiomas and Vascula Malformations of the Head and Neck. Toronto, Canada, John Wiley & Sons, 1999.

Hemangiomas are classified as benign vascular neoplasms and can be subdivided histopathologically into numerous categories depending on the literature. The most common histopathologic subdivisions include cavernous, capillary, and venous.[7] Although hemangiomas largely occur in the soft tissue, they have been documented to occur in intraosseous fashion in areas including but not limited to the vertebrae, skull, and facial bones. Cavernous hemangiomas are most commonly located in the vertebrae; however, they also can be located in the skull and even less commonly in the facial bones, as was noted in this patient.[2] Of the cavernous hemangiomas located in the skull, the most common site of these lesions is the frontal bone followed by the parietal bone.[1]

Intraosseous hemangiomas are benign, slowly expansile lesions that are most prevalent in women in the fourth and fifth decades of life.3, 6 These lesions are most likely traumatic or congenital in origin and symptoms vary depending on location.[8] Given their slowly expansile nature, when associated with the facial or skull bones they are often associated with a gradual worsening of nonspecific pain described as increased sinus pressure or even a dull headache as reported by this patient. However, these lesions have been associated with more severe findings, such as proptosis, loss of vision, and numbness.[9]

The differential diagnosis of these lesions often includes fibrous dysplasia, osteoid osteoma, eosinophilic granuloma, and osteosarcoma.[10] Intraosseous hemangiomas can have a “sunburst” or “honeycomb” appearance at imaging, as in the present case, that is not specific to their identity, thus warranting further workup to isolate a diagnosis.7, 9 Although a diagnosis cannot be made solely on imaging, there are some key findings to help differentiate intraosseous hemangiomas from fibrous dysplasia and osteosarcomas. Intraosseous hemangiomas have a sunburst or honeycomb appearance on CT, whereas fibrous dysplasia has more of a “ground-glass” appearance. Although osteosarcomas also can have a similar sunburst appearance on CT, they also will have a characteristic “onion skinning” reaction of the periosteum that is not typically found in intraosseous hemangiomas.[1] Preoperative imaging studies include conventional radiography, CT, and MRI. CT is the most useful imaging modality

owing to the ability of the imaging to capture cortical and trabecular details, such as the sunburst or honeycomb patterns. In contrast, MRI is used primarily to show depth of the tumor in relation to the soft tissue.[11]

Depending on the size and site, a biopsy can be performed to confirm the diagnosis. In this case, the lesion was small and in an esthetic zone of the face, negating that option. Once a diagnosis is made, treatment is usually patient specific, although a review of the literature suggests that angiography with en bloc surgical excision of the lesion is the treatment of choice.

On macroscopic examination, hemangiomas appear brown to dark red. They stand out as well-demarcated lesions of the medullary bone. Rarely, they can be intracortical or subperiosteal. Soft hemorrhagic cavities are crisscrossed with a honeycomb of fine trabeculae. Subperiosteal lesions can produce radiating spicules of bone at the surface.3, 6 On microscopic examination of carefully decalcified tissue, intraosseous hemangiomas are composed of large, dilated, endothelium-lined channels of varying size separated by loose to myxoid connective tissue (cavernous hemangiomas). Residual cancellous bone trabeculae are seen. The vascular channels are frequently engorged with red cells. Thrombus formation and calcification also can be seen occasionally.9, 10 The endothelial cells are flat and bland with no mitotic activity. Intraosseous capillary hemangiomas are exceedingly rare in bone, with only 5 cases documented in the head and neck region.9, 10

The authors conducted an extensive review of the literature on intraosseous hemangiomas of the frontal bone using EMBASE, PubMed, and Google Scholar. The search yielded 51 patients with an intraosseous hemangioma located in, or including a part of, the frontal bone from 1894 to December 2016. The patients had a mean age of 37.6 years, and 31 patients (~60%) were women. Most patients were asymptomatic at initial presentation, with their chief concern being the asymmetry or swelling caused by the lesion. The remainder of these patients presented with a headache or localized tenderness overlying the lesion. The workup focused on clinical examination and imaging, which included radiography, CT, or MRI or any combination of these modalities.

Intraosseous hemangiomas located in the frontal bone do not have a predilection for laterality. Lesions located in the frontal bone ranged from 1 to 6 cm, whereas some parietal and occipital lesions documented were as large as 8 cm. Intraosseous hemangiomas have been documented as occurring in all bones of the cranial vault and in bones of the face, including the orbit, nose, zygoma, maxilla, and mandible. Although the size of these lesions has been documented to grow as large as 8 cm, intraosseous hemangiomas of the nose rarely grow larger than 2 cm.

The most common treatment was preoperative angiography with embolization followed by en bloc resection of the lesion with or without reconstruction. Although the literature does not specify the recommended amount of free bony margins needed for en bloc resection, studies have shown successful removal without recurrence with free margins as small as 0.5 cm. Five patients (~10%) underwent partial resection of the lesion, and 1 of the 5 patients returned for a second procedure to excise the entire lesion. Four patients (~8%) chose nonsurgical options ranging from unspecified conservative treatment or no treatment at all.

Although many of these patients underwent en bloc resection of their intraosseous hemangioma, the options for reconstruction seem to vary. Options for reconstruction included split calvarial graft,

placement of titanium mesh with or without bone graft, bone cement, and polymethyl methacrylate cranioplasty. To the authors' knowledge, the present patient is the first documented case to use a porous polyethylene implant to immediately restore the defect stemming from en bloc resection of an intraosseous hemangioma in the frontal bone.

As noted earlier, there are limited data available in reference to reconstructive options after resection of intraosseous hemangiomas. The present patient underwent angiography and successful en bloc resection of a left frontal intraosseous hemangioma with subsequent placement of a porous polyethylene implant through the coronal approach. Her postoperative course was uncomplicated and there is currently no evidence of recurrence. She has healed appropriately with little to no complications and was pleased with the cosmetic result.

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