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Vascular malformation of the sphenoid and temporal bone: A diagnostic dilemma☆

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ABSTRACT

We present a rare case of a vascular anomaly of the sphenoid and temporal bones causing an expandable mass of the temporal region with dependent patient positioning and characteristic osseous changes on imaging. Initial diagnosis considerations included multiple myeloma (MM), fibrous dysplasia (FD), Paget's disease, lymphoma, meningoencephalocele (MEC), and vascular malformation (VaM). VaMs of the head and neck are rare and typically arise in the mandible and maxilla. However, this case demonstrates a unique finding of a VaM of the sphenoid and temporal bones with important radiological features to distinguish the diagnosis of vascular anomaly from other etiologies.

1. Introduction

Soft tissue vascular lesions of the head and neck can form a diagnostic challenge both clinically and pathologically. The landmark study by Mulliken and Glowacki developed a classification system for hemangiomas and VaM [1]. VaMs can include a single or combination of channels including capillary, lymphatic, venous, or arterial [2]. In the head and neck, the mandible and maxilla are the most common bones that are effected by congenital hemangiomas and VaM [3–5]. Intraosseous hemangiomas (IH) have also been described in the parietal and frontal bones, and less commonly, the zygoma, mandible and maxilla. However, IH of the skull base and orbit are exceedingly rare [3,6–8]. Diagnostic imaging can guide management of vascular lesions of the head and neck as there are characteristic diagnostic findings on computed tomography (CT) and magnetic resonance imaging (MRI) [3,9–13]. Skeletal changes associated with VaMs within the head and neck which have been previously described include distortion, erosion, hypertrophy, hypoplasia, and density changes from mechanical effects or primary intraosseous involvement [2]. We present a patient with an IH versus venous malformation (VM) arising from the sphenoid and temporal bones causing thickening and sclerosis as well as an expandable mass of the temporal region on physical exam.

2. Report of case

Brain MR imaging of a 76-year old female performed for evaluation of right hand tremor revealed an incidental 4.6 cm heterogenous, enhancing, trans-spacial mass involving the right squamous temporal bone, greater sphenoid wing, and overlying temporalis musculature (Fig. 1, A–C). Subsequent CT head performed a month later revealed a corresponding heterogenous radiolucent lesion involving the right sphenoid and temporal bones with cortical thinning (Fig. 3, A–B). Initial imaging differential considerations for a radiolucent lesion involving multiple bones of the face initially included Paget's disease, FD, venolymphatic malformation (VLM), MEC, MM, or lymphoma.

She underwent open biopsy in the operating room and final pathology revealed benign skeletal muscle, blood and fibrin without pathologic process. She was seen by neurosurgery at our institution who felt this lesion was consistent with FD with recommendation for continued observation with repeat CT scan in a year. A year after initial

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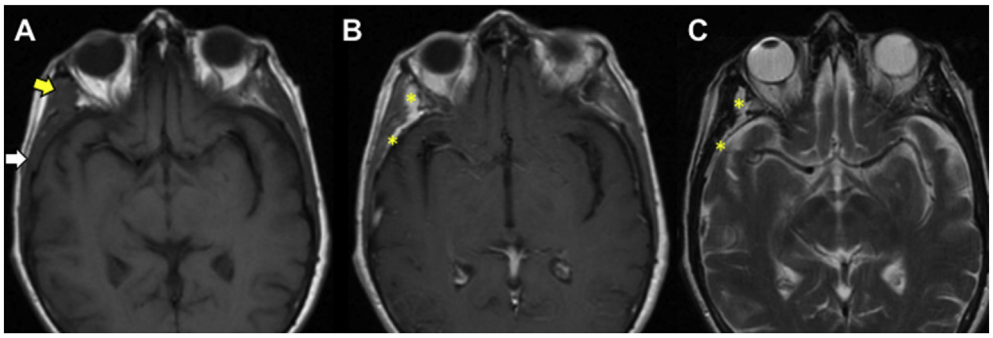

Fig. 1. Precontrast axial T1-weighted MR of the head (A) demonstrates a heterogenous multispacial lesion involving the right squamous temporal bone, overlying temporalis musculature (white arrow) and the right sphenoid triangle (yellow arrow). Axial T1-weighted postcontrast image (B) demonstrates heterogenous intralesional enhancement (yellow asterisks). Axial T2-weighted noncontrast image (C) demonstrates inhomogenous increased T2 signal within the lesion (yellow asterisks). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

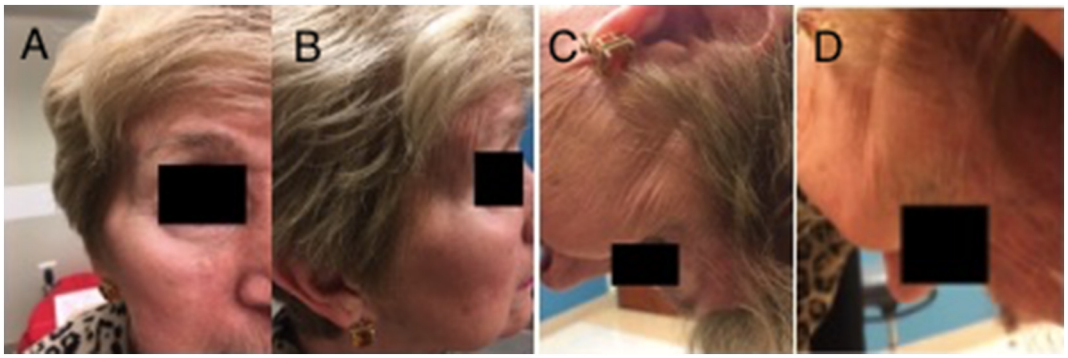

Fig. 2. Patient in upright position with depression of the right temporal area (A–B) and the expandable mass of the right temple while bending downward (C–D).

diagnosis, she noticed increased swelling of her right temple when bending down and looking to the left which immediately resolved when she stood up (Fig. 2, A–D). There were no associated visual changes, vertigo, headaches, or other neurological symptoms. The patient was referred to otolaryngology-head and neck surgery clinic for evaluation of right temple swelling, presumably attributable to the right-sided calvarial lesion.

She underwent right decubitus CT head to examine the expandable mass for evaluation of MEC, which demonstrated no interval progression of the lucent lesion involving the temporal and sphenoid bones. There was no evidence of meningocele with decubitus positioning and no osseous dehiscence the of MEC (Fig. 3, C). Review of the initial MR and CT imaging and subsequent CT imaging obtained approximately one year later at our institution was most suggestive of a VaM; IH versus VM. After discussion with the patient, she elected to observe clinically with serial imaging rather than medical management with sclerosing agent, repeat biopsy, or surgical removal.

3. Discussion

VaM of the sphenoid and temporal bones are exceedingly rare [6–8,14,15], and to our knowledge this is the first presentation of a VaM of the extracranial skull base without visual or neurological sequela. Though the lesion was initially suspected to represent fibrous dysplasia, pathology was not consistent with this diagnosis and demonstrated only normal skeletal muscle with no associated osseous abnormality. Additionally, the provocation with dependent head positioning is more consistent with a flow dependent lesion such as a VaM or MEC.

FD is a benign pathologic process caused by abnormal differentiation of osteoblasts where immature bone and fibrous stroma replace normal medullary bone [16]. The most craniofacial bones most commonly involved include the mandible and maxilla. The frontal, ethmoid, and sphenoid are less commonly involved and involvement of the temporal and occipital involvement with fibrous dysplasia is exceedingly rare [16]. Fibrous dysplasia has a characteristic appearance on CT, typically expansile with so called “ground glass” osseous matrix. On MRI, FD demonstrates a nonspecific appearance with heterogenous

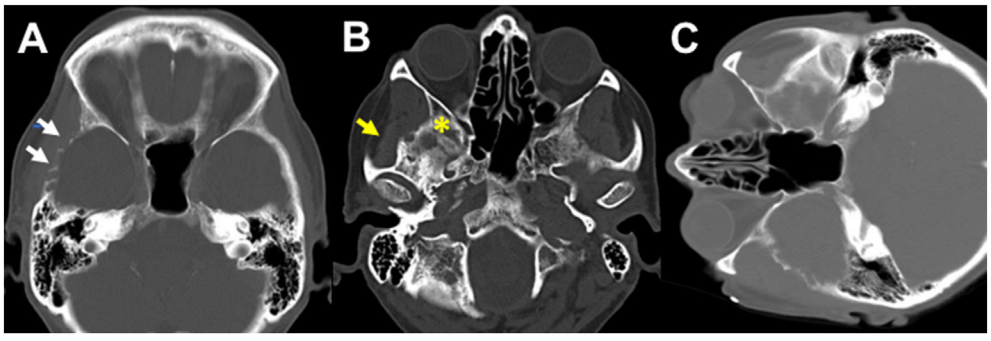

Fig. 3. Axial noncontrast CT (A,B) obtained 8 months after the initial MR exam demonstrates multifocal lucency and thinning involving the right squamous temporal bone (white arrows), right sphenoid triangle (yellow asterisk) and middle cranial fossa with thickening of overlying right temporalis muscle (yellow arrow). Left lateral decubitus noncontrast CT head obtained 9 months later (C) demonstrates no significant change in the lucency and thinning of the right temporal bone and sphenoid triangle. Specifically, no interval marrow expansion or further erosion was present. Lateral decubitus positioning reveal a meningocele. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

signal on both T1-weighted and T2-weighted images and heterogenous enhancement [16]. In our case, the appearance was not compatible with fibrous dysplasia as due to the lack of osseous expansion and lack of ground glass matrix.

Flow dependent expansion of the mass raised concern for MEC. Salehian et al. published on association of VM with MEC due to the presence of a VM causing a congenital error in development of the skull base [10]. No skull base defect was demonstrated in our patient, either by MR or on decubitus CT imaging. Additionally, MM and lymphoma were considered. However, lack of lesion progression, benign pathologic biopsy results, and lack of systemic symptoms suggestive of underlying malignancy were not compatible with the aforementioned diagnostic considerations.

IH, VM and arteriovenous malformations (AVM), have characteristics findings on CT and MRI which help to distinguish them. Hemangiomas, venous, capillary, and lymphatic malformations are considered low-flow lesions, whereas AVMs are considered high flow [13]. Hemangiomas have both a proliferative phase and an involutional phase [11]. Lui et al. reported a primary intraosseous skull base cavernous sinus hemangioma arising from the medial sphenoid wing causing chronic headaches and described the CT showing a mass causing abnormal mottled and trabeculated bone due to mechanical forces and primary intraosseous involvement [6]. MRI of hemangiomas often show lobulated masses that vary in heterogeneity with isointense signal on T1 to muscle and moderately intense signal on T2. However, hemangiomas can appear hyperintense on T2-weighted imaging relative to skeletal muscle, but lower signal that cerebrospinal fluid [9,13]. Asymptomatic VM are congenital malformations that are slow growing and can go unrecognized for decades. In particular, soft tissue VMs can present as masses that expand with dependence and “deflate” with standing [12]. On CT, soft tissue lesions can appear as a dynamically enhancing focal or trans-spatial mass while bony VMs can appear as a honeycomb expansive mass, but both are non-infiltrating. On MRI, VMs appear isointense on T1 and hyperintense on T2-weighted imaging. Generally, phleboliths and vascular lakes are highly suggestive of VM [9,12,13]. AVMs are considered high-flow vascular lesions which can present with bruits on physical exam. AVMs have a nidus, feeding vessels and large draining veins [11] which cause erosion and lysis of the bone on CT [9]. On MR imaging, AVMs demonstrate variable enhancement and may demonstrate flow voids on T2-weighted images [9,11,12]. Conventional angiography is frequently necessary for evaluation of AVMs, particularly when embolization is considered, in order to precisely map the feeding arteries and draining veins.

Our patient appeared to have a VaM favoring IH versus VM rather than FD due to lack of characteristic findings of FD (lack of expansile “ground glass” bone), the dependent filling of the lesion, edema of the overlying temporalis muscle, and thinning of the calvarium. On MRI, the lesion demonstrated variable enhancement with some hyperintense signal on T2-weighted images, consistent with VaM.

Without a definite histologic tissue diagnosis or angiography, it is difficult to definitively determine exactly what subtype of VaM our patient had. The type of vascular anomaly is an important factor when guiding management. Hemangiomas only need to be treated in the involution phase unless causing life threatening or visual disturbances [11]. Treatment of hemangiomas and VMs includes conservative management with observation and lifestyle modification, such as avoiding Valsalva maneuvers, medical management with beta blockers, steroids, or intralesional sclerotherapy, laser therapy, or surgery [6,11,12,15]. AVMs have a higher risk of hemorrhage and are typically treated with embolization and surgical excision [11,12,14]. Given our patient's age, the extracranial location, and lack of visual or neurological symptoms, we have elected to observe the lesion with clinical exam and serial imaging.

4. Conclusions

VaMs of the head and neck are congenital anomalies that rarely present on the sphenoid and temporal bone, frequently asymptomatic and discovered incidentally as in our patient. VaM should be considered in longstanding, lucent lesions, particularly in dynamic lesions which demonstrate expansion with provocative maneuvers (dependent head positioning in our patient).

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