Incidental cystic schwannoma of the left sphenoid and cavernous sinuses in an asymptomatic woman

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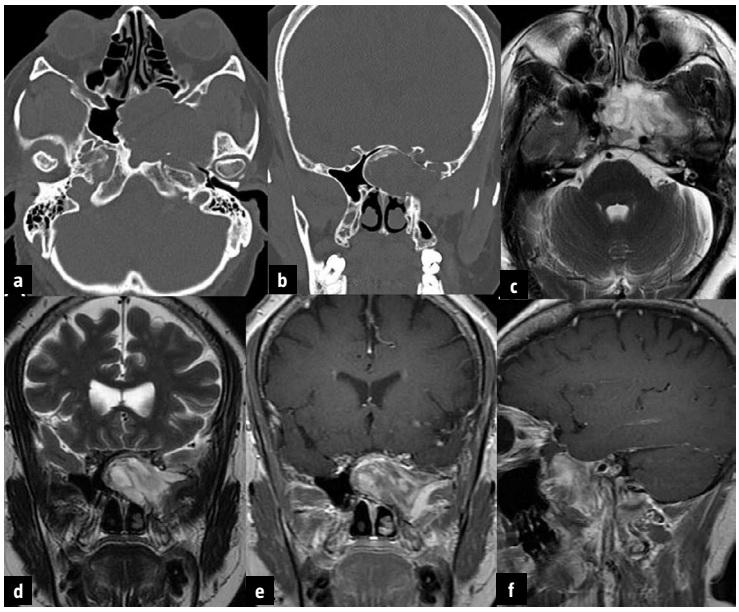

Figure. Computed tomoghraphy (a, b) axial and coronal planes revealed a large expansile bone defect with epicenter in the left sphenoid and cavernous sinuses. Magnetic resonance imaging (c, d) T_2 W axial and coronal planes revealed an irregular high-intensity space occupying process involving the sphenoid sinus and left cavernous sinus with extension into the left infratemporal fossa and masticator space. Magnetic resonance imaging (e, f) T_1 W post-contrast, coronal and sagittal planes respectively, demonstrated inhomogeneous enhancement of the described space occupying process.

Case report. A 63-year-old woman with a past medical history of hypertension and recent sinus infection presented to our institution due to an episode of syncope. She reported no sinus pressure or pain, changes in vision, headache, or weight loss. Physical and neurological examinations were both normal, including no cranial nerve deficits. As part of her workup she underwent a computed tomography scan of the head (Figure, a, b), which incidentally discovered a large 3.8 × 3.6 × 2.4 cm bone defect by an expansile mass with apparent epicenter in the left sphenoid sinus with adjacent remodeling of the sphenoid sinus, clivus, greater wing of the sphenoid bone, and tip of the left petrous bone. There was no obvious evidence of brain extension. Brain magnetic resonance imaging with and without contrast (Figure, c-f) demonstrated a 4.8 × 3.4 × 2.1 cm expansile, complex, partially cystic mass apparently arising from the lateral sphenoid/cavernous sinus with extension to the medial aspect of the left middle cranial fossa, masticator space, and cavernous sinus. The mass demonstrated heterogenous enhancement on contrast images. No hemorrhagic component was seen on gradient-recalled echo. A biopsy was performed with the impression of a large mucocele, with frozen pathology showing nuclear atypia. Immunohistochemistry showed spindle cells that were positive for S-100 and vimentin. Final pathology report stated the left sphenoid mass was a schwannoma. Our patient underwent endoscopic sinus surgery with image guidance for debulking of her mass. Her original syncope was ultimately deemed to be of vasovagal etiology.

Discussion. Schwannomas, also referred to as neurilemomas, are benign nerve sheath tumors of Schwann cells that can arise anywhere throughout the body, with the head and neck making up for 25-45% of cases [1]. Of these head and neck neoplasms, only 4% arise in the sinonasal region [2]. Reports of the literature consistently identify the sphenoid sinus as a most uncommon location for sinonasal schwannomas [1]. Schwannomas have a characteristic radiographic appearance, commonly appearing as ovoid well-circumscribed soft tissue masses that preserve most of their bony margins [2]. As these tumors enlarge, cystic and hemorrhagic degeneration become more characteristic features [2]. These tumors are defined on magnetic resonance imaging by hypointensity on T_1, hyperintensity on T_2, and significant enhancement following contrast [2]. Pathologically, schwannomas are characterized by spindle cells with wavy nuclei, and a positive immunohistochemistry staining for S100 protein and vimentin, and negative staining for epithelial and smooth muscle markers [3].

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Only three other reports of cystic schwannomas arising exclusively from the sphenoid sinus have been documented in the literature [1,3,4]. Symptomatology in the previously described cases was varied, although none were completely asymptomatic [1,3,4]. DiNardo and Mellis reported a 60-year-old man with a 2-month history of diplopia on lateral gaze with decreased visual acuity, headache, and hyposmia [1]. Dutta et al reported a 22-year-old man with a 6-month history of headaches and progressive painless unilateral vision loss [3]. Amri et al reported an 85-year-old man with a 3-month history of right nasal obstruction [4]. Our patient, however, was completely asymptomatic despite her tumor's large size and involvement of the left sphenoid and cavernous sinuses, and presents as the first asymptomatic report of this rare neoplasm.

Of note is that in three of these four cases, including ours, the initial diagnosis was thought to be of mucocele [1,3,4]. Paranasal sinus mucoceles are inflammatory lesions that can also present with an array of symptomatology, ranging from asymptomatic, rhinological, neurologic, and ophthalmologic [5]. Mucoceles can also mimic cystic schwannomas on radiologic imaging, as these cystic lesions can appear initially hypointense on T_1 and hyperintense on T_2 [5]. As mucoceles age, their T_1 intensity increases, T_2 intensity diminishes, and they become more associated with bony remodeling [5]. Ultimately, given the ambiguity of the clinical picture painted by a cystic schwannoma in the area of the sphenoid and cavernous sinuses, the final diagnosis must be made via biopsy and immunopathologic studies.

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