Pterygopalatine Fossa Mucocele: A Common Paranasal Sinus Mass in an Uncommon Location

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The pterygopalatine fossa (PPF) is a complex 3-dimensional subsite of the skull base located behind the maxillary sinus. Within the PPF are numerous neural and vascular structures that connect the nasal cavity, orbit, middle cranial fossa, and infratemporal fossa.¹ Primary tumors of the PPF are rare and derive from the nerves or blood vessels within this space.² Mucoceles can develop in the paranasal sinuses due to trauma, prior surgery, or severe infection.³ Because mucosa is not located within the PPF, mucoceles are generally not considered in the differential diagnosis for a PPF mass. Here we describe the occurrence, radiographic diagnostic dilemma, and successful surgical management of an asymptomatic mucocele within the PPF.

Case Report

This study was deemed exempt from review by the Department of Research Programs at Walter Reed National Military Medical Center.

The patient is a 22-year-old African American woman with a history significant for craniosynostosis, bilateral cleft lip and palate, and midface hypoplasia. She had undergone numerous head and neck surgeries, including cleft repair, bilateral Le Fort III osteotomies, and craniosynostosis repair. Computed tomography (CT) of the temporal bone was performed for conductive hearing loss, showing hardware and distorted anatomy from prior surgery. It also found an incidental $1.3 \times 1.6 \times 1.9$ -cm expansile, soft tissue mass in the left PPF without destruction of the surrounding bone (Figure 1). A benign nerve sheath tumor or encephalocele was considered the most likely diagnosis. Magnetic resonance imaging (MRI) of the face/orbits demonstrated a nonenhancing soft tissue mass with similar T1 and T2 signal intensity to brain parenchyma, further suggesting an encephalocele vs a purely cystic schwannoma with proteinaceous fluid contents (Figure 2). She was otherwise asymptomatic and denied facial hypoesthesia, nasal congestion, rhinorrhea, or vision issues.



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In clinic, endoscopic evaluation was unremarkable. Treatment options were discussed, and she elected to proceed with endoscopic exploration and biopsy of the mass. Endoscopic sinus surgery, including left maxillary antrostomy and septoplasty, was performed. The left PPF was explored after the posterior wall of the maxillary sinus was taken down. The mass was visualized and entered with sharp dissection, and the mucocele was drained with an immediate return of thick nonpurulent mucus. The mucocele was marsupialized, and the cavity was inspected. There was no cerebrospinal fluid leak. The postoperative course was uneventful.

Discussion

Masses of the PPF typically derive from the normal structures within the PPF. The most common masses include juvenile nasopharyngeal angiofibromas (JNAs), schwannomas, or lipomas. Masses may also breach the PPF, including encephaloceles or malignancy.^{2,3} Imaging is critical in the workup and diagnosis of PPF lesions and can assist operative planning. JNAs frequently show PPF widening, anterior bowing of the posterior wall of the maxillary sinus (Holman-Miller sign), and bony remodeling on CT. On MRI, JNAs are heterogeneous on T1 and T2 with hypervascular flow voids and display intense enhancement with contrast.⁴ Schwannomas

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Figure I. Computed tomography of the head in soft tissue window (A) and bone window (B) reveals ovoid soft tissue mass expanding the left pterygopalatine fossa. Anteriorly, hardware and distorted anatomy seen from prior surgery.



Figure 2. Magnetic resonance imaging of the face with axial TI-weighted (A) and T2-weighted (B) images confirms a mass in the left pterygopalatine fossa (arrow) with similar signal intensity to the left temporal lobe.

typically show a well-defined mass on CT that remodels the surrounding bone as it grows. On MRI, schwannomas tend to be isointense on T1, hyperintense on T2, and enhancing with contrast.⁴ Lipomas are a well-defined soft tissue mass with fatty attenuation (negative Hounsfield units) on CT. On MRI, lipomas are expected to follow fatty signal on all sequences (hyperintense on T1). Encephaloceles show a smooth lesion extending through an associated skull or skull base defect on CT. On MRI, they demonstrate herniation of often abnormal brain tissue and surrounding CSF/meninges through the defect.⁴

Mucoceles found in the paranasal sinuses present as expansile lesions without bony destruction on CT. On MRI, they typically show low signal intensity on T1 and high signal intensity on T2, although there can be high variability depending on the proportion of water, mucus, and protein.⁴ For example, higher protein content accounts for the higher T1 and lower T2 signal in this case, somewhat mimicking brain tissue. Although it is impossible to confirm which surgery or event may have caused the mucocele in this patient, we hypothesize that mucosa was introduced and subsequently trapped in the PPF from the patient's Le Fort III osteotomy for midface hypoplasia, since others have described mucocele formation in the infratemporal fossa after prior endoscopic sinonasal surgery or prior trauma with midface fractures.^{3,5} In conclusion, clinicians should consider mucoceles in the differential diagnosis for patients with a history of orthognathic surgery who present with an expansile nonenhancing PPF mass.

Author Contributions

Robert J. Lewis, conducted literature review and wrote the manuscript; **Robert Y. Shih**, reviewed and edited manuscript for content; **Anthony M. Tolisano**, generated project idea, edited the manuscript; **Charles A. Riley**, generated project idea and edited the manuscript for content.

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