# A Rare Cause of Sphenoid Sinusitis

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**Abstract**

Malignancy of the sphenoid sinus is rare. Tumors may extend to adjacent anatomic structures before detection and may be easily missed. Outcomes are typically poor and vary by tumor type. Clinicians should maintain vigilance for neoplastic disease in patients presenting with headache and ocular/neurological complaints of the face/sinuses.

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A Rare Cause of Sphenoid Sinusitis

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Key Clinical Message (Abstract):

Malignancy of the sphenoid sinus is rare. Tumors may extend to adjacent anatomic structures before detection and may be easily missed. Outcomes are typically poor and vary by tumor type. Clinicians should maintain vigilance for neoplastic disease in patients presenting with headache and ocular/neurological complaints of the face/sinuses.

Key Words: lymphoma, headache, sphenoid sinus

Introduction:

Malignancy of the sphenoid sinus is rare, representing less than one percent of all cancers [1]. The most common types are squamous cell, adenocarcinoma, non-Hodgkin's mature B-cell lymphoma, and unspecified epithelial neoplasms [1]. Tumors may extend to adjacent anatomic sites before detection [2]. Each tumor may present with a broad range of symptoms, such as chronic sinusitis, and may be easily missed [3]. Outcomes

are typically poor [1]. Survival rates, however, vary by type of tumor [4]. The most common age group affected is 50-59, predominantly white males [1]. Average tumor size is 3.7cm [1].

Case:

A 54-year-old white male complained of an intermittent, increasingly severe, headache for two weeks. On the day of presentation to the emergency department, he awakened at 0600 with increasingly severe right periorbital pain, described as sharp, radiating down his right cheek and the right side of his neck. He had two episodes of nausea/vomiting. He denied any syncope, visual changes, vertigo, slurred speech, extremity weakness/paresthesias, fever/chills, or trauma. Review of systems was otherwise negative. Past medical history includes diabetes mellitus, migraine headaches, and Meniere's disease. He denied and tobacco, alcohol, or drug use. Family history includes migraines. On exam the patient was awake/alert with stable vital signs. Patient was afebrile. HEENT exam revealed pupils to be equal/round/reactive to light. Photophobia was noted in the right eye. Extraocular muscles intact. No cranial nerve deficits. No nasal drainage. Tenderness over the right temporal artery noted. Uvula midline. No tongue deviation on protrusion. No evidence of carotid bruits noted. No cervical adenopathy or nuchal rigidity. The remainder of the exam was unremarkable. Diagnostic results revealed a white cell count of 4.0 and an ESR of 2. Lumbar puncture was negative. CT of the head revealed complete opacification of the sphenoid sinus. CT angiogram of the head revealed no abnormalities. Patient was admitted for further work-up, antibiotics, and ENT consultation. Patient initially refused an MRI, however, he ultimately consented to an MRI of the brain which was negative. MRA demonstrated stenosis of the right anterior cerebral artery and right middle cerebral artery. Two days later he developed a third nerve palsy and diplopia in the right eye secondary to optic nerve involvement. Biopsy of the sphenoid sinus revealed a large B-cell lymphoma, which was wrapped around the optic nerve.

Patient treatment consisted of six months of chemotherapy with negative PET scans on follow-up. Approximately five years thereafter the patient was diagnosed with a GI bleed, fatty liver, and esophageal varices (for which banding was completed). Further evaluation revealed malignant hepatic lesions and metastasis to the spine causing back pain. Palliative radiotherapy was initiated but the patient passed shortly thereafter prior to further work-up of the hepatic lesions.

Discussion:

Primary B-cell lymphoma of the sphenoid sinus is very rare, having an incidence of 0.03 per 100,000 persons between 2000-2012 [1,2,5,6]. B-cell lymphomas remain the most common non-epithelial tumors of the paranasal sinuses [2,7]. The sphenoid sinus, contained within the confines of the sphenoid bone, is anatomically contiguous with the carotid arteries, optic nerves, the maxillary division of cranial nerve V, brain stem, and sella turcica [3]. Consequently, clinical presentations vary widely [5,7]. Headaches, often in the temporal, retroorbital, and retrobulbar regions or, cranial neuropathies, and facial pain/paresthesias may all occur [3,8,9,10]. Headaches are thought to be secondary to tumor effect on the sphenopalatine ganglion and remain the most common presenting symptom [4]. The anatomic location makes resection challenging due to adjacent structures [1,4]. Imaging often reveals opacification of the sphenoid sinus [4,5]. CT and MRI remain the imaging modalities of choice [10,11]. Fortunately, B-cell lymphomas are rapidly-growing and thus very sensitive to chemotherapy and radiotherapy [5]. Surgery may be possible in select cases [1,4]. Prophylactic intrathecal chemotherapy may be utilized as there is a potential for recurrence in the CNS [6]. Biopsy of sphenoid sinus opacification must be considered in chronic sinusitis or when other previously mentioned signs/symptoms occur [5,10,12,13]. Clinicians should maintain a high degree of vigilance for neoplastic disease in patients presenting with headache and ocular/neurological complaints of the face/sinuses.

Ethical Statement:

I certify that this material has not been published elsewhere, either in whole or part, and is not under consideration for publication in any other journal. I have personally and actively been involved in substantive work leading to the revised manuscript and the authors will hold themselves jointly and individually responsible for its content.

Statement of Acknowledgement:

This case report was published with the written informed consent of the patient's wife.

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