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Chondromyxoid fibroma involving the sphenoid sinus: Case report and literature review

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We present the case of a 62-year-old woman with chondromyxoid fibroma of the sphenoid sinus. Chondromyxoid fibroma is a rare bone tumor found most prevalently in long bones, so its presence at the cranial base is especially uncommon. The presence of a monoclonal gammopathy of undermined significance (MGUS) prompted consideration and investigation of a plasma cell disorder; however, CT and MRI findings followed by biopsy led to the correct diagnosis of chondromyxoid fibroma.

Introduction

Chondromyxoid fibroma is a benign cartilaginous neoplasm (1) first distinguished from other cartilaginous tumors by Jaffe and Lichenstein in 1948 (1, 2). It is exceedingly rare, accounting for 0.5% of the 10,065 bone tumors categorized by Unni and Inwards and 1.6% of their catalog of benign bone tumors. Only 2 of the 50 chondromyxoid fibromas included in their study occurred in the skull. In another study of 76 cases of chondromyxoid fibroma including additional cases from the literature, 1 of 189 were in the skull (3), and in a review of 278 cases, 15 were in the skull or facial bones (4). Chondromyxoid fibroma consists of lobulated areas of interspersed myxoid, fibrous and chondroid material (1). This benign neoplasm occurs most frequently in young adults, and it is found in numerous anatomic locations, including long bones, flat bones, and cranio-facial bones (1).

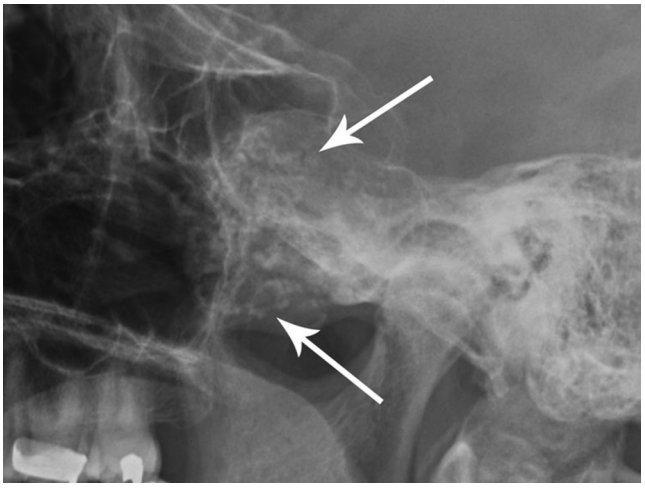

Figure 1. 62-year-old woman with chondrdomyxoid fibroma involving the sphenoid sinus. Lateral view of the skull. A mass arising from the skull base partly fills the sphenoid sinus and contains calcifications, many with rounded, "rings-and-arcs" configuration (arrows).

Case report

A 62-year-old woman was found, on workup for incidental right leg pain, to have an elevated serum total protein due to a small MGUS. A bone survey ordered as part of the evaluation of possible plasma cell dyscrasia revealed a lytic lesion at the base of the skull with heterogeneous calcifications, some having ring-and-arc configuration (Fig. 1). Other laboratory tests and marrow aspiration showed no evidence of multiple myeloma. Nonetheless, the presence of

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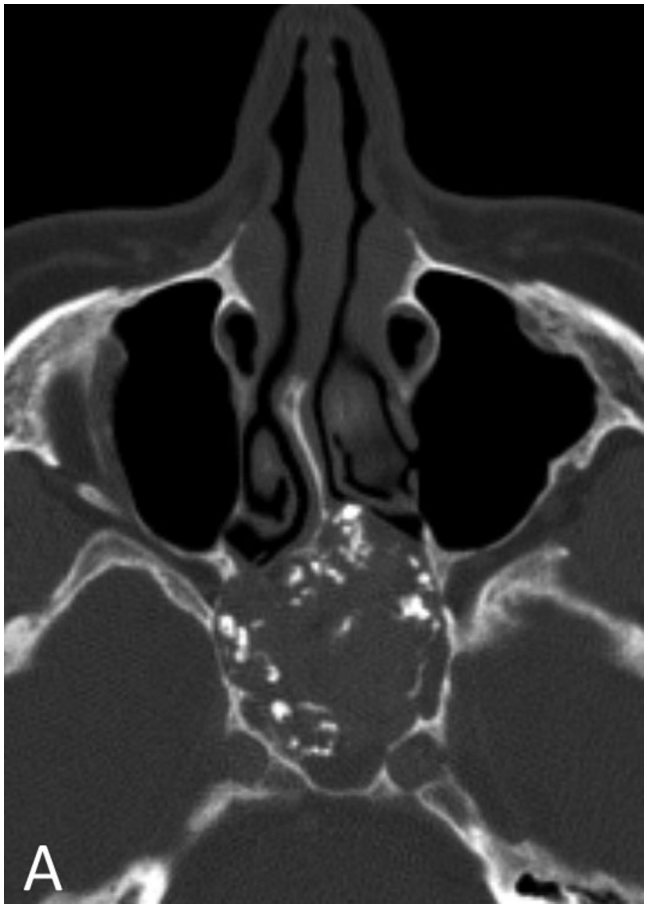

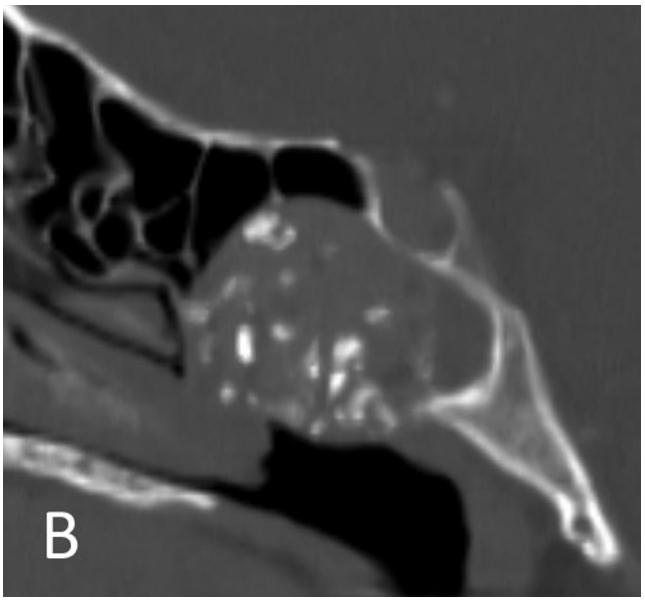

Figure 2. 62-year-old woman with chondrdomyxoid fibroma involving the sphenoid sinus. Axial (A) and sagittal (B) CT images demonstrate a mass with coarse calcifications just caudal to the sella turcica. Axially, images show an expansile midline mass with smoothly expanded, benign bony margins. On the sagittal image, the mass extends into the posterior aspect of the nasal cavity and superior aspect of the nasopharynx. Bony margins are harder to evaluate on this view, but the clivus does not appear to be eroded.

MGUS raised the question of a solitary plasmacytoma or amyloidoma.

The patient had prior ocular complaints including right orbital pain and blurred vision, recurrent corneal erosion, and bilateral cataract surgery. There were no symptoms clearly related to the bone lesion.

A CT scan for further evaluation of the skull lesion (Fig. 2, A and B) confirmed a 33 x 33 x 33-mm mass arising in the central base of the skull with coarse calcification. In addition to plasmacytoma and amyloidoma, chondrosarcoma and chordoma were among the diagnostic considerations. Ultimately, a transsphenoidal biopsy revealed a chondromyxoid fibroma.

An MRI scan was performed after biopsy to evaluate the residual mass and to serve as a baseline for follow-up (Fig. 3). The plan was to perform pre-operative embolization and surgical resection in case the tumor had grown. So far, after two years, serial MRI studies have shown stability. The patient remains asymptomatic with regard to the lesion, and there has been no change in level of the small MGUS.

Discussion

Chondromyxoid fibroma is a rare cartilage-producing benign tumor, accounting for 1.6% of benign tumors in the series studied by Unni and Inwards (1). Chondromyxoid fibroma is often a round or oval-shaped lytic lesion less than 5 cm in diameter, usually found in the metaphysis of long tubular bones, but occasionally involving the base of the skull, mandible, frontal bone, or nasal bone. Chondromyxoid fibroma usually presents in early adulthood (1).

The rarity of chondromyxoid fibromas involving any part of the skull means that they should never occupy the primary place in the differential diagnosis of any skull lesion, even one that ultimately turns out to be a chondromyxoid fibroma. The differential diagnosis for a well-circumscribed lesion with chondroid-like matrix calcification such as in this patient would include other more frequent chondroid lesions, particularly chondrosarcoma and chordoma (5).

Chondromyxoid fibroma appears similar to these other cartilage tumors on radiography, CT, and MRI. All demonstrate decreased signal on T1-weighted images and heterogeneous increased signal on T2-weighted images. On radiography and CT, they often present with calcification of the chondroid matrix (6, 7). Heterogeneity on T2-weighted images is due to varying chondroid, myxoid, and fibrous elements throughout the tumor. With gadolinium, all of these lesions will usually enhance (6, 7). Chondromyxoid fibromas will usually have well-circumscribed borders (1).

Chondrosarcomas and chordomas often both exhibit frank bone destruction (6, 7). Typically, chordomas are thought to occur in the midline, and chondrosarcomas are considered to occur off midline in the area of the petro-

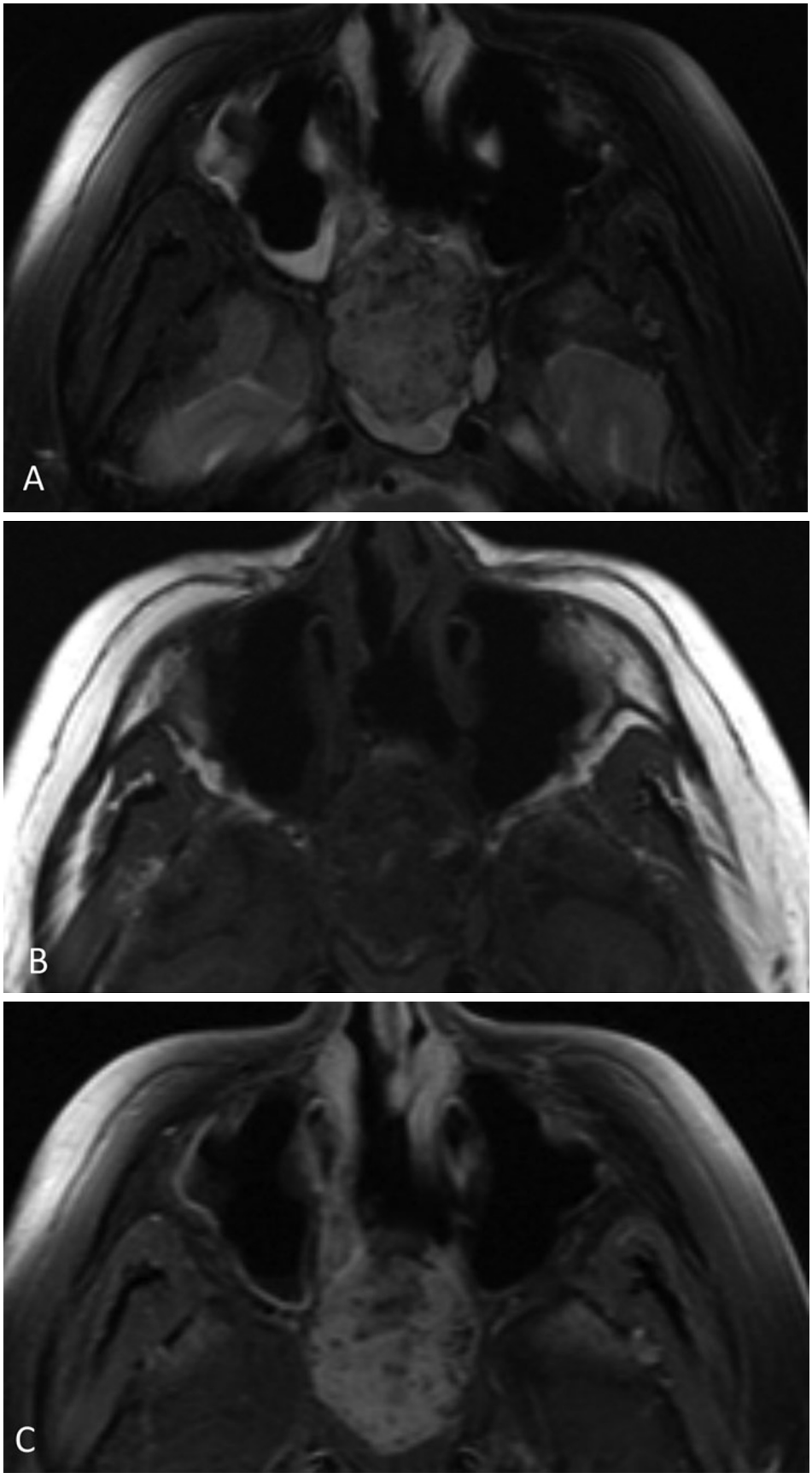

Figure 3A-C.62-year-old woman with chondrdomyxoid fibroma involving the sphenoid sinus. Axial T2, T1, and postcontrast T1-weighted MR images through the mass at the base of the skull confirm that the majority of the tumor remains intact after biopsy. Areas of signal dropout correspond to the coarse calcifications seen on conventional radiography and on CT scan. The mass is hyperintense on T2 and appears noninfiltrative. It enhances after contrast administration.

occipital fissure (7). A recent study of 38 chordomas and 4 low-grade chondrosarcomas occurring in the base of the skull did not, however, confirm this dogma (6).

For the lesion described in this report, additional items in the differential diagnosis were plasmacytoma and amyloidoma, as suggested by the presence of MGUS, which may be associated with various plasma-cell disorders, including solitary plasmacytoma of bone, multiple myeloma, primary amyloidosis, and amyloidosis with myeloma (8). Amyloidosis involves the abnormal accumulation and deposit of amyloid proteins in tissues. Amyloidomas at the skull base are very rare. Since osseous amyloidomas often contain coarse calcifications, they may be mistaken for cartilage tumors, particularly chondrosarcoma (9, 10). However, a lower signal on T2-weighting, approximately that of skeletal muscle, rather than hyperintensity, is a common feature of amyloidoma (11, 12). Although a bright signal on T2-weighting is the rule for plasmacytomas and focal lesions of multiple myeloma elsewhere in the body, those at the base of the skull have, like amyloidomas, been reported to be relatively low in signal on T2-weighted images (13).

Chondromyxoid fibroma is often treated via excision (14). However, postexcision recurrence is common, as the tumor may not

be removed completely and may recur (15-17). Such recurrence is usually local, and malignant conversion is unlikely (14). Radiation therapy, however, is generally avoided due to reported cases of malignant transformation (7). In our patient, followup rather than excision was chosen because the patient was asymptomatic and because of the anticipated complexity of resection.

This patient's history included MGUS with a differential diagnosis that therefore included amyloidoma and plasmacytoma. Biopsy was necessary for proper diagnosis and rational planning of long-term followup.

The table on the final page summarizes information on eight known cases of chondromyxoid fibroma involving the sphenoid sinus, including ours.

References

1. Unni K, Inwards C. *Dahlin's bone tumors*. Lippincott Williams & Wilkins; 2010.
2. Jaffe HL, Lichenstein L. Chondromyxoid fibroma of bone: a distinctive benign tumor likely to be mistaken especially for chondrosarcoma. *Arch Pathol*. 1948; 45:541-51. [PubMed]
3. Rahimi A, Beabout JW, Ivins JC, Dahlin DC. Chondromyxoid fibroma: a clinicopathologic study of 76 cases. *Cancer*. 1972; 30:726-736. [PubMed]
4. Wu CT, Inwards CY, O'Laughlin S, Rock MG, Beabout JW, Unni KK. Chondromyxoid fibroma of bone: a clinicopathologic review of 278 cases. *Hum Pathol*. 1998; 29(5): 438-46 [PubMed]
5. Keel SB, Bhan AK, Liebsch NJ, et al. Chondromyxoid fibroma of the skull base: a tumor which may be confused with chordoma and chondrosarcoma: a report of three cases and review of the literature. *Am J Surg Pathol*. 1997; 21:577-82. [PubMed]
6. Pamir MN, Özduman K. Analysis of radiological features relative to histopathology in 42 skull-base chordomas and chondrosarcomas. *Eur J Radiol*. 2006 Jun; 58(3): 461-70. Epub 2006 May 2. [PubMed]
7. Otto BA, Jacob A, Klein MJ, Welling DB. Chondromyxoid fibroma of the temporal bone: case report and review of the literature. *Ann Otol Rhinol Laryngol*. 2007 Dec; 116(12): 922-7. [PubMed]
8. Kyle RA. The monoclonal gammopathies. *Clin Chem*. 1994 Nov; 40(11 Pt 2): 2154-61. [PubMed]
9. Ünal F, Hepgül K, Bayindir C, Bilge T, Imer M, Turantan I. Skull base amyloidoma. Case report. *J Neurosurg*. 1992 Feb; 76(2): 303-6. [PubMed]
10. Simoens WA, van den Hauwe L, Van Hedent E, et al. Amyloidoma of the skull base. *AJNR Am J Neuroradiol*. 2000 Sep; 21(8);1559-62. [PubMed]
11. Oruckapten H, Karli Oğuz K, Isikay I, Ruacan S. Amyloidoma of the temporal bone and upper cervical spine; presentation of a rare clinical entity with a brief literature review. *Turkish Neurosurgery*. 2009; 19(2):159-162. [PubMed]
12. Gean-Martin AD, Kirsch CF, Vezina LG, Weber AL. Focal amyloidosis of the head and neck: evaluation with CT and MR imaging. *Radiol*. 1991; 181:521-525. [PubMed]
13. Cerase A, Tarantino A, Gozzetti A, Muccio CF, Gennari P, Monti L, Di Blasi A, Venturi C. Intracranial involvement in plasmacytomas and multiple myeloma: a pictorial essay. *Neuroradiology*. 2008 Aug; 50(8): 665-74, Epub 2008 May 31. [PubMed]
14. Patino-Cordoba JI, Turner J, McCarthy SW, Fagan P. Chondromyxoid fibroma of the skull base. *Otolaryngol Head Neck Surg*. 1998 Mar; 118(3 Pt 1): 415-8. [PubMed]
15. Mirra JM. *Bone tumors: clinical, radiologic, and pathologic correlations*. Philadelphia: Lea and Febiger; 1989: 648-8.
16. Zillmer DA, Dorfman HD. Chondromyxoid fibroma of bone: thirty-six cases with clinicopathologic correlation. *Hum Pathol*. 1989 Oct; 20(10): 952-64. [PubMed]
17. Shek TW, Peh WC, Leung G. Chondromyxoid fibroma of skull base: a tumour prone to local recurrence. *J Laryngol Otol*. 1999 Apr; 113(4): 380-5. [PubMed]
18. Frank E, Deruaz JP, de Tribolet N. Chondromyxoid fibroma of the petrous-sphenoid junction. *Surg Neurol*. 1987; 27: 182-6. [PubMed]
19. Nazeer T, Ro JY, Varma DG, de la Hermosa JR, Ayala AG. Chondromyxoid fibroma of paranasal sinuses: report of two cases presenting with nasal obstruction. *Skeletal Radiol*. 1996; 25: 779-782. [PubMed]
20. Yu E, Vescan A, Perez-Ordonez B, Lazinski D, Bartlett E. Chondromyxoid fibroma of the skull base: case report and review of the literature. *J Otolaryngol Head Neck Surg*. 2009 Oct; 38(5): E114-7. [PubMed]
21. Vernon SE, Casiano RR. Sphenoid sinus chondromyxoid fibroma mimicking a mucocele. *Am J Otolaryngol*. 2006; 27: 406-8. [PubMed]
22. Morris LG, Rihani J, Lebowitz RA, Wang BY. Chondromyxoid fibroma of sphenoid sinus with unusual calcifications: case report with literature review. *Head and Neck Pathol*. 2009 Jun; 3(2): 169-173. Epub 2009 Jun 10. [PubMed]; PubMed Central PMCID: PMC2715466

Cases	Authors	Age/Sex	Location	Borders	Calcified	MRI	Expansile	Treatment	Followup
1	Frank/18	26/m	Petrous/ sphenoid bone	Well-circumscribed	Yes, by conventional radiographs and CT	Not obtained	Yes, into the clinoid process, sella, cavernous sinus, and retrosellar area	Complete surgical removal	NA
2	Nazeer/19	66/f	Sphenoid sinus	Not specified	Not specified	T1 isointense, T2 hyperintense, enhanced with gadolinium	Yes, into nasopharynx and sella	Surgery	Local recurrence after 1 yr; curetted, 6 mos FOD
3	Keel/5	65/f	Sphenoid/ occipital bone	Well-circumscribed	Imaging was not reported	Imaging was not reported	Yes, involved the clivus (where it was thought to have originated), sphenoid sinus, & ethmoid sinus	Surgery	26 months FOD
4	Keel/5	66/f	Sphenoid/ occipital bone	Well-circumscribed	Imaging was not reported	Imaging was not reported	Yes, into the ethmoid sinus & nasopharynx	Surgery and radiation	Local recurrence after 6 mos; after radiation, 20 mos FOD
5	Yu/20	39/m	Sphenoid sinus - temporal mandibular joint	Infiltrative	None by CT	T1 intermediate signal, T2 predominantly high signal	Yes, involved the left middle cranial fossa (from which it was considered to have arisen), cavernous sinus, sphenoid sinus, masticator space, temporomandiblar joint	Surgery	6 months stable MRI
6	Vernon/21	43/m	Sphenoid sinus	Well-circumscribed	No, the tumor resembled a mucocele	Obtained but not described	Yes, into the nasopharynx	Surgery	FOD
7	Morris/22	52/f	Sphenoid sinus	Well-circumscribed	Yes, by CT	Not obtained	Yes, into the nasal cavity	Surgery	2 years FOD
8	Haygood/ this case	62/f	Sphenoid sinus	Well-circumscribed	Yes, by conventional radiographs and CT	T1 intermediate signal, T1 predominantly high signal, enhanced with gadolinium	Yes, into the nasal passages & nasopharynx	Biopsy for diagnosis, then observation	2 years stable MRI

NA: Not applicable
FOD: Free of disease