**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).

**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 the 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.
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-
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
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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.

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| 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 retrostellar 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; curedtted, 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, temporomandibular 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