Aggressive Metastatic Chordoma in the Wrist

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Sir:

Chordoma is a rare, malignant, primary bone tumor (1–4), which arises from remnant cells of the developmental notochord.[1] We report an extremely rare case of a metastatic sacral chordoma in the wrist.

A 59-year-old man presented in 2012 with a gradually enlarging 3 cm bulbous, transilluminating, firm mass over the left volar radial wrist. The mass was suspicious for a ganglion cyst and was aspirated at an outside institution with indeterminate pathology (Fig. 1). The patient had a history significant for a 14×12 cm sacral chordoma, which was surgically excised in 2011, with a total sacrectomy and pelvic reconstruction and had been disease free for 1 year. An excisional biopsy was performed (Fig. 2), and pathologic analysis revealed a 3×3 cm metastatic myxoid chordoma positive for brachyury. Magnetic resonance imaging of the left wrist for staging, 2 months after initial diagnostic excision, demonstrated a recurrent 1.3×0.7 cm lobulated mass. A second resection was performed with a 1-cm wide margin. In 2013, patient developed recurrence iliac and lumbar vertebrae disease treated with Gleevec. The wrist is now 15 months post-op and is without signs of local recurrence, but patient continues to demonstrate vertebral metastases. There are no further surgical interventions planned for the patient.

The differential diagnosis of masses frequently identified in the hand includes ganglion cysts, lipomas, tendon sheath giant cell tumors, schwannomas, neurofibromas, fibromatoses, sarcomas, and metastases.[2] The classical morphology of chordomas has characteristic vacuolated physalipherous cells in a myxofibrillary background separated into clusters/lobules.[1,3] Chordomas are differentiated from histologically similar tumors via immunological markers, which include S-100, EMA, CK-10 and 19, and most recently brachyury.[3,4]

Chordomas are extremely rare with an annual incidence of 0.0001% in the general population, with approximately 300 new patients diagnosed each year in the United States.[5] Chordomas are slow growing, locally aggressive, and typically found at the

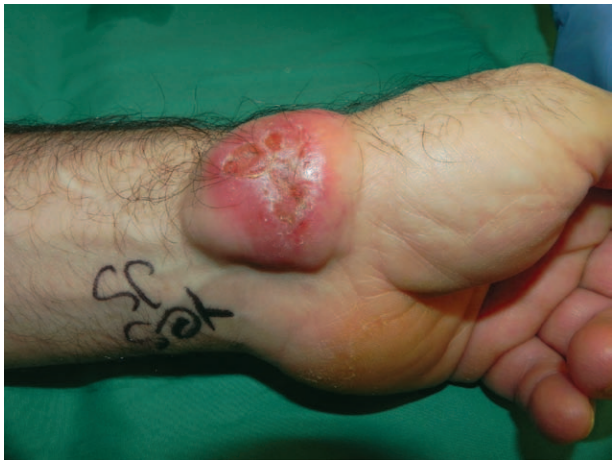

Fig. 1. Preoperative view of a 59-year-old man with a gradually enlarging left volar radial wrist mass.

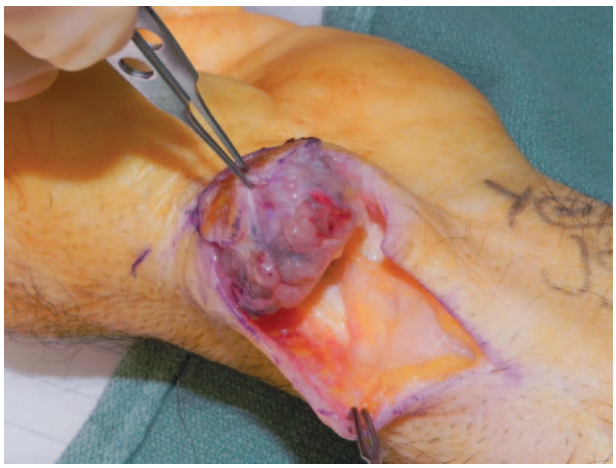

Fig. 2. Intraoperative view of excisional biopsy. Note the clean separation of the vascularized myxoid tumor from the underlying soft tissue without evidence of invasion of the deep fascia.

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skull base and sacrococcygeal region. Median age of presentation is reported to range from 59 to 62 years, with a gender predilection in sacrococcygeal chordomas for men of 2:1.[5] Clinical presentation is dependent on the location of the enlarging mass, with sacral tumors presenting with chronic lower back pain, lower extremity neurological deficits, and bowel/bladder dysfunction while intracranial tumors presenting with headache, diplopia, and craniofacial pain. Primary treatment for a chordoma is wide margin surgical excision.[1] Local recurrence rates greater than 70% have been reported in surgically treated chordomas with nonnegative margins, and although chordoma metastasis is rarely evident during initial diagnosis, it presents later in the disease process in 40–60% of patients.[1,5] Common sites for metastasis include the lungs, liver, heart, lymph nodes, and vertebrae.[5]

Metastatic chordomas present a significant diagnostic challenge due to the rarity of the primary tumor and the nonspecific presentation of the metastatic mass. This case illustrates the need for increased physician vigilance in monitoring newly identified masses in patients with a history of chordoma, by obtaining a biopsy and subsequent surgical excision of the metastatic mass.

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DISCLOSURE

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