In the middle of the nineteenth century, Rudolf Virchow first described small tumors with round, physaliphorous (bubblelike) cells in the region of the clivus, whose origin he erroneously assumed lay in the synchondrosis sphenooccipitalis. In 1858, Muller described the similarity of these tumors to the notochord. With this in mind he suggested the term “chordoma.” Chordomas are believed to arise from remnants of the notochord within the vertebral bodies, sacrum, and clivus.

The distribution of chordomas is 50% sacrococcygeal, 35% clival, and 15% in the remaining vertebral column. Chordomas account for between 1 and 3% of primary bone tumors. Overall these lesions are associated with a male predominance (male/female patient ratio 2:1) but this varies according to the site of the lesion, with the sacrococcygeal site being found more often among older women.

In 2000 the patient began to experience progressive confusion and impairment of higher cognitive functions. A CT scan of the brain, obtained at that time, demonstrated a lobular tumor with surrounding vasogenic edema in the right occipital lobe (Fig. 3).

The patient was placed on a regimen of steroid medications and there was some clinical improvement. We obtained CT scans of the thorax, abdomen, and pelvis, and the findings were normal.

Operation. The patient underwent a right-sided occipital craniotomy and total excision of the mass. The lesion was well circumscribed and adherent to the dura mater. The entire tumor as well as the involved dura was resected.

Postoperative Course and Pathological Findings. The patient made a good postoperative recovery. Histopathological studies showed a chordoma identical to the primary neoplasm in the sacrum (Fig. 4).

In 2003 the patient was found to have a large recurrence of the sacral chordoma. Partial tumor excision was performed. Magnetic resonance images of the brain did not reveal any recurrence of the cerebral lesion (Fig. 3 lower left and right).

Currently, the patient is confined to a wheelchair because of the sacral lesion. There is no evidence of any other distant metastasis.

Discussion

Chordoma consists of physaliphorous cells, which resemble cartilage. In some cases, highly malignant areas with cellular pleomorphism, hyperchromatism, and increased mitotic activity similar to sarcoma are observed. Metastasis may occur in these cases. The occurrence

**Intracranial metastasis from a sacrococcygeal chordoma**

**Case report**


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Chordoma is a locally invasive tumor of low metastatic potential. Only six cases of chordoma that metastasized to the brain are found in the English literature. Most of these lesions were clinically silent and all were associated with extraneural metastases. The authors report a case of symptomatic brain metastasis from a sacrococcygeal chordoma in the absence of other metastases. The incidence, sites, and factors predictive of chordoma metastasis are discussed.

KEY WORDS • chordoma metastasis • brain metastasis

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**Case Report**

History. This 69-year-old man underwent a posterior partial sacrectomy and debulking of a sacral chordoma in 1998, followed by a temporary defunctioning colostomy (Figs. 1 and 2). He also underwent postoperative local radiotherapy.

Examination. A 2-year follow-up observation of the residual sacral mass demonstrated that it remained stable in size.

Abbreviation used in this paper: CT = computerized tomography.
Intracranial metastasis from a sacrococcygeal chordoma

The mean age of patients with proven metastasis from chordoma is 45 years in men and 37.3 years in women. According to Higinbotham, et al., and Chambers and Schwinn, metastases have been observed more frequently with primary localized vertebral chordomas than with sacrococcygeal chordomas. Only a small number of metastases from clival chordomas have been described. These results may reflect the relatively poorer prognosis associated with clival chordomas.

In 76% of chordomas that subsequently metastasized, the primary tumor had been irradiated, probably reflecting its incomplete removal. Incomplete excision may allow residual tumor to gain access to the bloodstream and predispose to metastasis.

Chambers and Schwinn have pointed out that most of the 70 cases of metastatic chordoma in their review were incidentally discovered only at autopsy. They conclude that metastatic disease in such cases is usually subclinical. Most cases of metastasis present with symptoms and signs referable to the primary lesion or to disease recurrence at the primary site following therapy.

The present case is unusual in three respects. First, chordomas rarely metastasize and when they do, it is usually to the lung, lymph nodes, or bone. The brain is an extremely rare site for metastasis. Second, in none of the six cases of chordoma metastatic to the brain reported in the literature was metastasis confined only to the brain. Third, the presentation in this case was one of a symptomatic metastasis (rather than of symptomatic recurrence at the primary site), which is a rare phenomenon.
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