

CLINICAL CASE

Isolated liver metastasis from sacral chordoma. Case report and review of the literature

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Summary

Chordoma is a rare malignant tumor derived from remnants of the primitive notochord. It can occur anywhere along the skull base and spine. The most commonly involved area is the sacrococcygeal region. Although chordomas invade adjacent structures, they metastasize less often. We

present a case of a sacrococcygeal chordoma with liver metastasis. The nature and origin of the liver lesion was proved with fine-needle aspiration (FNA) under computed tomographic (CT) guidance.

Keywords: chordoma, FNA, liver metastases, rare malignant tumor

Introduction

Chordomas constitute 3-4 % of all primary bone tumors [1-3]. They arise from remnants of the notochord and can occur anywhere along the skull base and spine. Fifty percent arise in the sacrum, 35% in the clivus and 15% in the vertebrae [1,3].

Although chordomas invade adjacent structures, they metastasize less often. However, chordomas arising in the vertebral bodies display a more malignant behavior than their counterparts in the sacrum or the clivus [1].

Case presentation

A 72-year-old woman with a known sacrococcygeal chordoma came to our hospital for her annual

follow up. The patient had undergone surgical excision of the tumor 6 years ago, followed by radiation therapy. The CT examination of the sacrococcygeal area revealed extended local recurrence (Figure 1), while that of the upper abdomen revealed a hypodense lesion with unsharp borders in the liver (segment VIII), measuring 3×2 cm and showing features of secondary deposit (Figure 2). No intravenous contrast material was administered due to her personal history of major allergic reaction. Therefore, we decided to proceed to FNA of the lesion under CT guidance (Figure 3).



Figure 1. Sacrococcygeal chordoma 6 years after the operation. Extended local recurrence with infiltration of the muscles (short arrow) and osteolytic lesion of the left ilium (long arrow).

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Figure 2. Hypodense round lesion with quite unsharp margins in the right lobe of the liver.

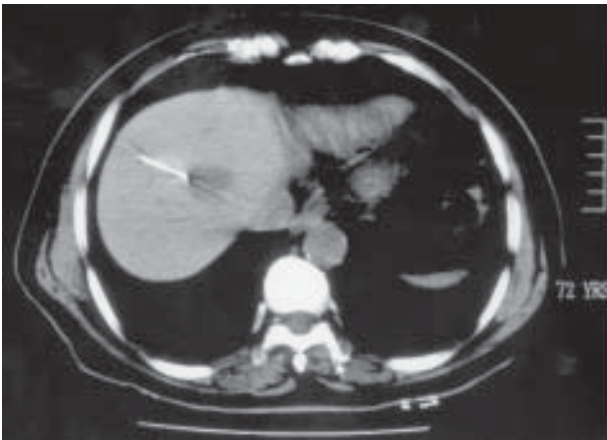


Figure 3. Fine-needle aspiration of the liver lesion under CT guidance.

The clinical and laboratory investigations were negative. Cytological evaluation proved that the above lesion was a metastatic focus from the known sacrococcygeal chordoma (Figures 4 and 5).

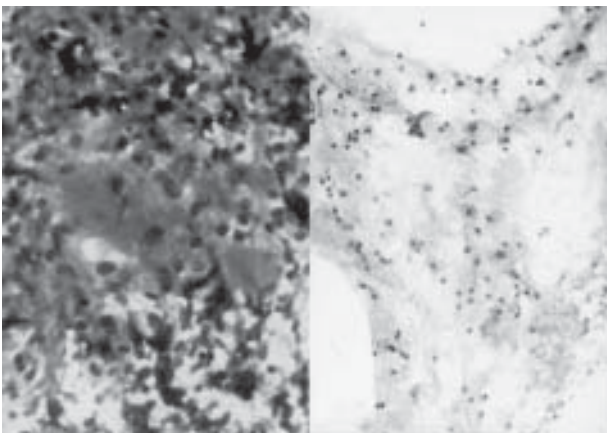


Figure 4. Vacuolated cells with mild nuclear pleomorphism lie within a myxoid stroma (MGG $\times 1200$ and Papanicolaou $\times 600$).

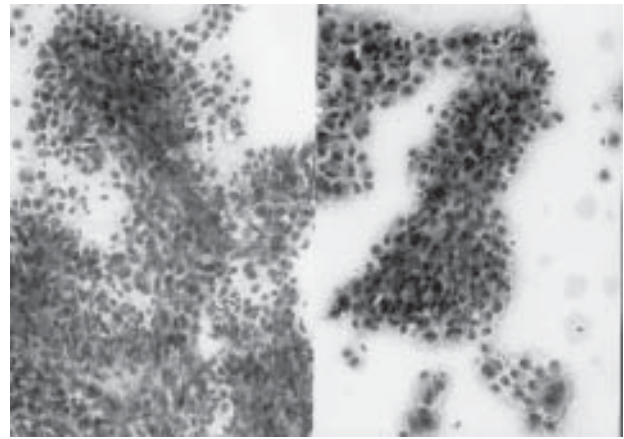


Figure 5. Purple myxoid background material like a network encircles the tumor cells (MGG $\times 600$).

Discussion

Chordoma is an infrequent tumor arising from remnants of the fetal notochord, which represents the earliest fetal axial skeleton extending from the Rathke pouch to the coccyx [1-4].

Chordoma occurs most commonly in people aged 30-60 years, with the peak incidence in the 5th decade [1,5]. Unlike sphenoid lesions which have an equal sex distribution, spinal chordomas have 2-3:1 male to female ratio [1,3,4].

The clinical presentation is often subtle, with a gradual onset of neurological symptoms including pain, numbness, motor weakness, and incontinence or constipation in cases with sacral localization.

There are 3 common sites of origin. Most common is the sacrum, second comes the region of the clivus and third the area of the upper cervical spine. The tumors are rather slow-growing and of low-grade malignancy, spreading by infiltration and metastasizing only in the late stages of the disease [1,3].

Grossly, chordomas may be soft, tan, myxoid masses that frequently show areas of hemorrhage [2].

Pathologically, these lobulated neoplasms are often contained within a pseudocapsule. Microscopically, the hallmarks of chordoma are the bubble-like or physaliferous cells with multivacuolated cytoplasm which is arranged in cords, columns, sheets or trabeculae within large lakes of extracellular mucoid matrix. Some cells may have eosinophilic non-vacuolated cytoplasm, while others contain large vacuoles imparting a signet ring appearance. Nuclei are usually mild with non-existent or rare mitotic figures, small clusters and single cells in a fibrillary myxoid background [2,4,6,7]. Chordomas are divided into 3 sub-

types based on microscopic morphology: a) conventional, b) chondroid, c) dedifferentiated [2].

The most frequent radiographic appearance of chordoma is that of a destructive lesion of a vertebral body centered in the midline, with a large, associated soft-tissue mass. Osseous expansion is frequent, and evidence of intratumoral calcification is seen in 50-70% of sacrococcygeal lesions on radiographs and in as many as in 90% on CT scans [1,8,9]. The calcification is typically amorphous and often predominates at the periphery of the lesion. Lesions of spinal segments above the sacrum are generally less expansible, have calcification in only approximately 30% of the cases, and may contain sclerosis (43-62% of the cases) [1,8]. In some cases sclerosis predominates, leading to an 'ivory' vertebral appearance. There may be intervertebral disk involvement, with narrowing of the disk space, which is unusual for most spinal tumors and simulates infection. This appearance has been described in 11-14% of chordomas and may lead to involvement of 2 adjacent vertebral levels [3,4,7]. Similarly, sacrococcygeal chordomas may extend across the sacroiliac joint.

CT demonstrates both the osseous and the soft-tissue components of chordoma and the affected surrounding structures. Coronal oblique CT of sacrococcygeal lesions is the optimal method for detecting neural foraminal and sacroiliac joint involvement. In more than 50% of cases, CT shows low attenuation within the soft-tissue mass, which implies the presence of myxoid-type tissue [8-10]. A higher-attenuation fibrous pseudocapsule is commonly seen at the periphery of the lesion.

Magnetic resonance imaging (MRI), because of its multiplanar capabilities and exquisite contrast resolution, is superior to CT in depicting the extent of chordoma [4,9]. Generally, chordoma has low to intermediate signal intensity on T1-weighted MR images and has very high signal intensity on T2-weighted images (similar to the nucleus pulposus). This characteristic reflects the high water content of these lesions. High signal intensity has occasionally been reported on T1-weighted MR images in intracranial or sphenococcygeal chordomas and is presumably related to the high protein content of the myxoid material. However, in our experience this MRI appearance is rare with spinal chordomas. Enhancement of chordoma is commonly seen after intravenous administration of contrast material, either at CT or MRI, similar to the vascular blush reported with angiographies [4,8-10].

The prognosis of chordoma depends on whether the tumor can be completely resected. The loca-

tion of the lesion and its very large size at presentation often necessitates incomplete excision. Adjuvant radiation therapy may also be employed in the treatment of chordoma. Generally, patients with sacrococcygeal lesions have an improved survival, possibly related to the ability to achieve a more complete resection, averaging 8-10 years, compared with 4-5 years for patients with chordomas at other sites [4]. Death is often related to local recurrence and invasion rather than to metastatic disease.

The reported prevalence of metastatic spread of chordoma in the literature varies widely, reaching even up to 40% according to some authors. Sites of metastases include the liver, lungs, regional lymph nodes, peritoneum, skin and heart, with no reports about the percent involvement of each organ [2,4,5,8].

The presented case is therefore unusual in two respects. Firstly, because, according to the relevant literature, the liver is a comparatively uncommon site for metastatic spread; and secondly, in our more than 15-year experience in our oncologic hospital, this is the first case of chordoma with liver metastasis.

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