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Humeral metastasis of sacrococcygeal chordoma detected by fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography: A case report

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ABSTRACT

Chordomas are rare, slow-growing, locally aggressive bone tumors arising from embryonic remnants of the notochord. Distant metastases most commonly involve the lung, liver, axial skeleton, skin, and lymph nodes. Humeral metastases are extremely rare. We report the case of a recurrent chordoma with humeral metastasis, complicated with pathologic fracture. Fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography revealed multiple hypermetabolic skeletal lesions, corresponding to the symptoms. Our report suggests that positron emission tomography-computed tomography is useful for evaluation of recurrence and distant metastases of chordomas.

Introduction

Chordomas are very rare tumors arising from the remnants of the notochord in the sacrum (50%), skull base (30%), and mobile spine (20%); These tumors are low-grade, locally invasive malignancies. The metastatic potential is relatively low. Distant metastases most frequently involve the lung, liver, axial skeleton, skin, and lymph nodes. However, metastases to the humerus are extremely rare. Metastases usually occur late in the natural course of the disease, mostly after local recurrence [1]. Whole-body fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography (^{18}F-FDG PET/CT) appears to be useful for evaluation of distant metastases with a single examination. Here we report a case of sacrococcygeal chordoma with humeral metastasis diagnosed by ^{18}F-FDG PET/CT.

Case report

An 84-year-old man presented to our hospital with right arm pain and right shoulder joint impaired mobility after minor activity. The man had a histopathologically proven sacrococcygeal chordoma 3 years ago and had been treated with surgical resection and postoperative radiotherapy.

Competing Interests: The authors have no personal, financial, or institutional interest with regard to the authorship and publication of this article.

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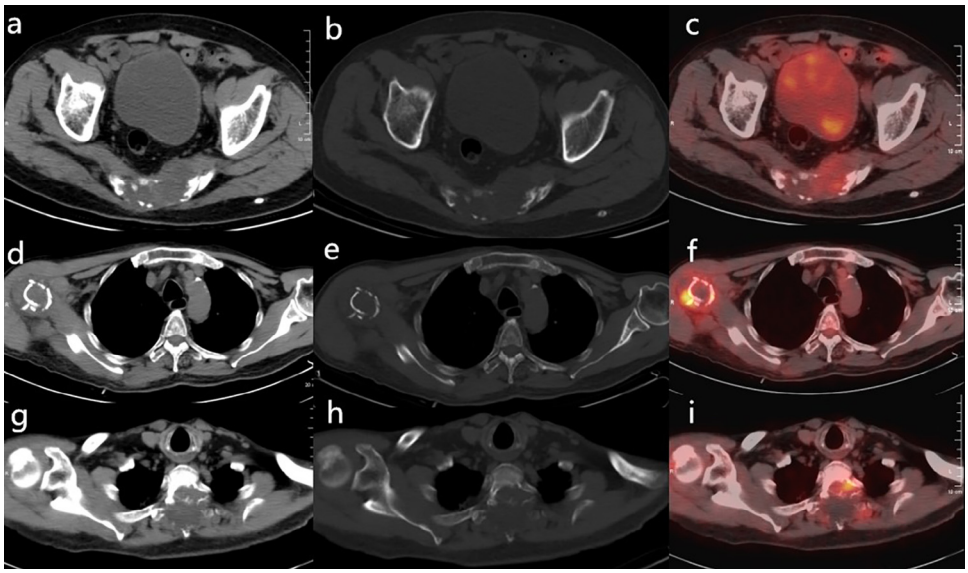

Fig. 1 – The fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography images showing multiple osteolytic destructive lesions with increased fluorine-18 fluorodeoxyglucose uptake in the sacrococcygeal region (A-C), the proximal shaft of the right humerus (D-F), and T2 and T3 (G-I).

The patient had ^{18}F-FDG PET/CT for restaging. Transaxial ^{18}F-FDG PET/CT showed an osteolytic destructive lesion with extraosseous extension in the sacrococcygeal region with increased tracer uptake, suggestive of local recurrence (Fig. 1A-C). There were also osteolytic destructive lesions in the proximal shaft of the right humerus and the second and third thoracic vertebrae (T2 and T3) with increased tracer uptake, suggestive of metastases (Fig. 1D-I). The patient was treated with surgery thereafter. Based on the histopathologic (Fig. 2), immunohistochemical, and imaging findings, the patient was diagnosed with recurrent sacrococcygeal chordoma with the right humeral and the T2 and T3 metastases. The immunohistochemical staining of the humeral lesion showed S-100 (+), phosphoenolpyruvate carboxykinase (+), epithelial membrane antigen (partly, +), vascular endothelial growth factor receptor 2 (–), vimentin (+), sex determining region Y (SRY)-related high mobility groupbox 9 (partly, +), integrase interactor 1 (–), CD117 (–), brachyury (–), and mindbomb E3 ubiquitin protein ligase 1 (–) (Fig. 3).

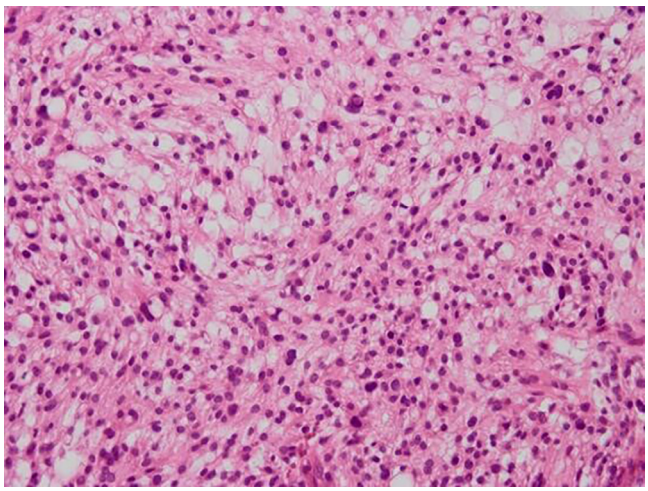

Fig. 2 – Biopsy of the humeral lesion showing cords and nests of epithelioid tumor cells in a background of extracellular chondroid material (hematoxylin and eosin, original magnification ×200).

Discussion

Chordomas are rare, low-grade, locally invasive primary bone tumors arising from the embryonic remnants of the notochord, around which the base of the skull and the vertebral column develop. Remnants of the notochord usually remain in or close to the midline. The anatomic distribution of the chordomas mirrors the location of notochord remnants. The sacrum is the most common site of origin, accounting for 50% of all cases, followed by the skull base (30%) and the mobile spine (20%) [1]. The median age at presentation is around 60 years. However, presentation with skull base tumors may occur in a younger population, including children [1–4]. Grossly, chordomas are soft, lobulated, semitranslucent, gray masses that often hemorrhage and permeate and destroy bones. Under the microscope, chordomas display lobules and vacuolated, moderately atypical neoplastic cells across a myxoid stroma separated by fibrous bands [4].

The reported incidence of metastatic disease of chordomas is 19%. Metastases usually occur late in the course of the disease. Tumors with local recurrence are more likely to develop metastatic disease, which mostly involves the sacrococcygeal region, lung, liver, axial skeleton, and lymph nodes [5,6]. Large tumor size, inadequate surgical margins, local