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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.

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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 (¹⁸F-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 ¹⁸F-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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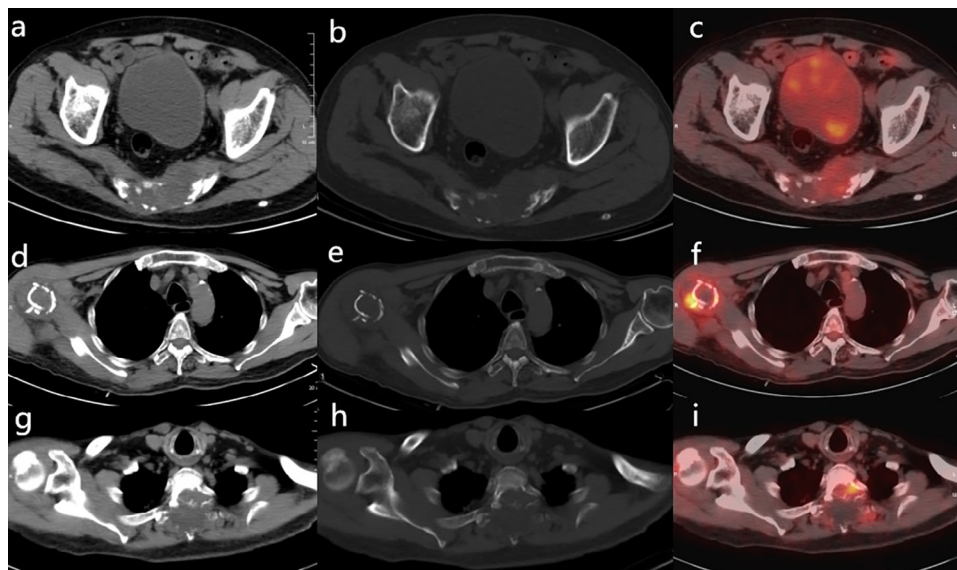


Fig. 1 – The fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography images showing multiple osteolytic destructive lesions with increased fluorine-18 fluorodeoxyglucose uptake in the sacrococcygeal region (A-C), the proximal shaft of the right humerus (D-F), and T2 and T3 (G-I).

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).

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

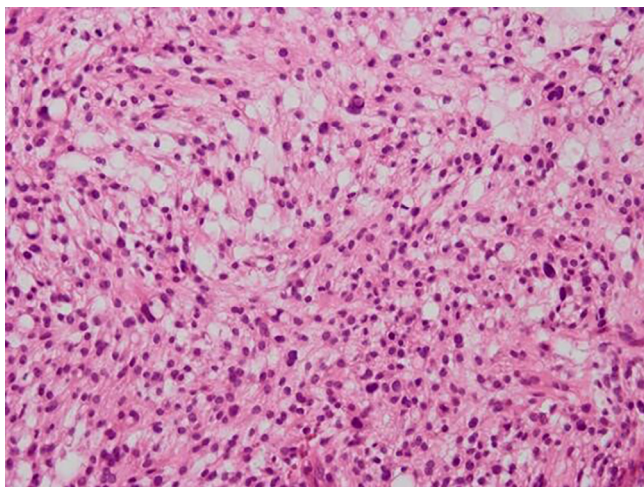


Fig. 2 – Biopsy of the humeral lesion showing cords and nests of epithelioid tumor cells in a background of extracellular chondroid material (hematoxylin and eosin, original magnification $\times 200$).

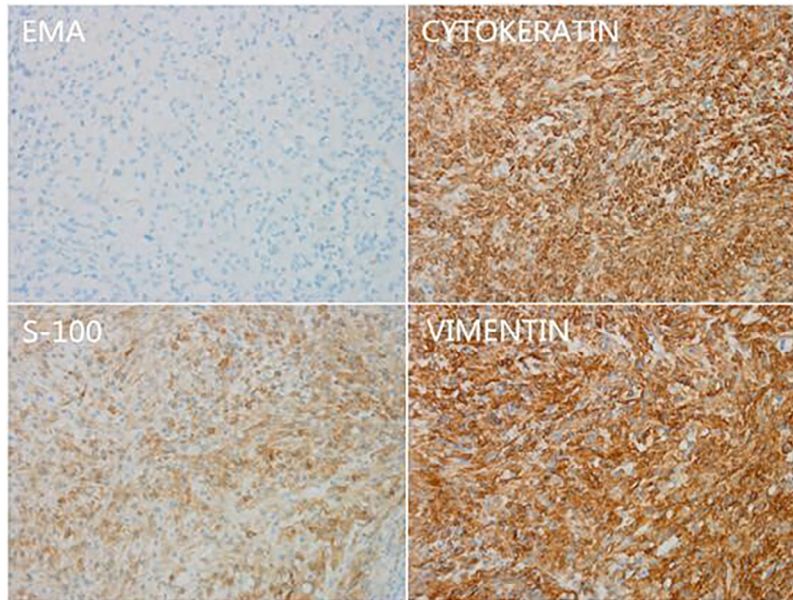


Fig. 3 – Immunohistochemistry demonstrating the tumor cells of the humeral lesion staining positive for EMA, cytokeratin, S-100 and vimentin, consistent with metastatic chordoma (hematoxylin and eosin, original magnification $\times 200$). EMA, epithelial membrane antigen.

aggressiveness, local recurrence, microscopic tumor necrosis, high Ki-67 labeling index, and invasive diagnostic procedure have been reported to be adverse prognostic factors [7,8].

Humeral metastasis of chordoma is rare. There have been only 2 reports, both originating in the sacrococcygeal region [9,10]. The common primary cancers that metastasize to the humerus are breast, lung, prostate, kidney, and thyroid carcinomas [11]. Chondrosarcomas, malignant fibrous histiocytomas, and osteosarcomas are relatively rare but should also be considered [11,12]. In our case, ^{18}F -FDG PET/CT is helpful in evaluating the local recurrence and metastatic disease of chordoma and in diagnosing metastatic disease based on the tracer uptake.

The therapeutic approach to chordoma has traditionally relied heavily on surgery. Indications for definitive radiation therapy are unresectable disease, inoperable patients, or neurologic impairment not accepted by the patient. Chordomas are usually not sensitive to chemotherapy except for high-grade dedifferentiated tumors [1–3]. Most chordomas strongly express epidermal growth factor receptor and platelet-derived growth factor receptors receptor, and some promising responses to the molecularly target agent imatinib have been reported [13].

Magnetic resonance imaging has been widely used for assessing tumor invasion, especially for central nervous system primary tumors and spinal metastases, because of clear delineation of lesion and adjacent tissue structures. Most chordomas show mild to moderate heterogeneous enhancement on contrast-enhanced magnetic resonance imaging scan. Computed tomography is necessary for surgical planning [1,2,14]. Whole-body ^{18}F -FDG PET/CT is useful for the assessment of chordomas with a single examination, especially when local recurrence or distant metastasis is suspected. Chordomas usually show heterogeneously increased tracer uptake on ^{18}F -FDG PET/CT scan [14–20].

Conclusion

We report a case of sacrococcygeal chordoma with humeral metastasis and T2 and T3 metastases. Although bone is one of the common metastatic sites of chordomas, metastasis to the humerus is extremely rare. ^{18}F -FDG PET/CT is useful for the detection and exact localization of local recurrence and distant metastasis of chordomas with a single examination.

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