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Case report

Gluteus maximus metastasis from sacrococcygeal chordoma: A case report [☆]

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ABSTRACT

We reported the metastatic chordoma in the right gluteus maximus of a 73-year-old man. The patient was initially diagnosed with sacrococcygeal chordoma and treated with surgical resection. Unfortunately, he had a gluteal metastasis and recurrence and suffered the third-operation therapy. Chordoma is a rare malignant tumor and may metastasize, but the metastasis and recurrence of chordoma at the right gluteus maximus following sacrococcygeal region are extraordinarily rare. This unusual case report may be helpful to clinical workers in diagnosing chordoma.

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Introduction

Chordoma is a rare malignant tumor originating from the remnants of the embryological notochord. The reported incidence of chordoma is 0.08/100000[1], ie, 0.1/100,000 in males and 0.06/100,000 in females[2,3]. In adults, chordoma is located typically in the sacrococcygeal (50%), the intra cranial at the skull base(35%), and intermediate spinal regions (15%).^[4] Chordoma has been recently reported to have a 5- and 10-year survival rates of 73%–86% and 49%–71%, respectively^[5]. The

most common location of metastatic chordoma is the lung followed by the liver, soft tissues, and bone^[6]. To date, we are unable to find other cases of chordoma metastases to the gluteus maximus in our search of the literature.

Case report

In 2018, a 73-year-old male patient with a history of sacrococcygeal chordoma surgically resected four years prior was

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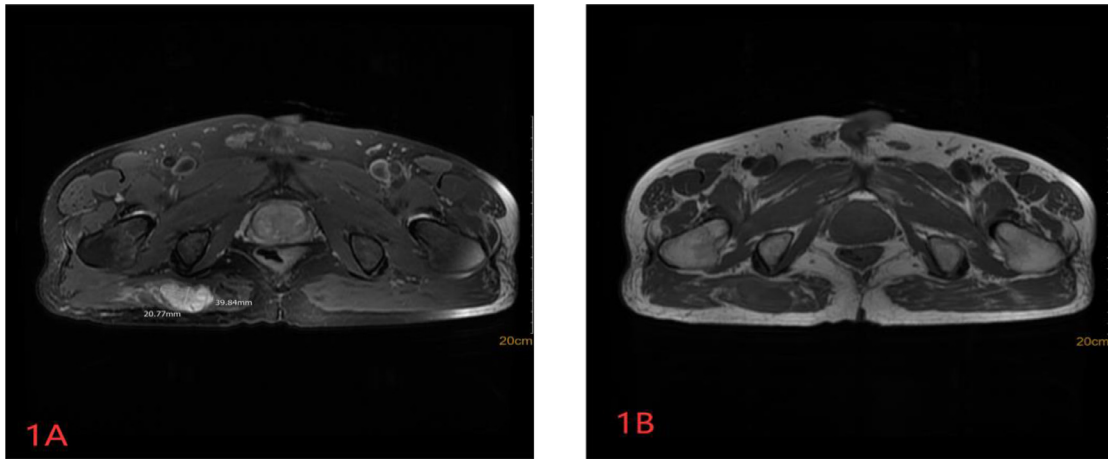


Fig. 1 – Large mass with hyperintense on T2-weighted images in the right gluteus maximus (A) and isointense on T1-weighted images (B) in 2018

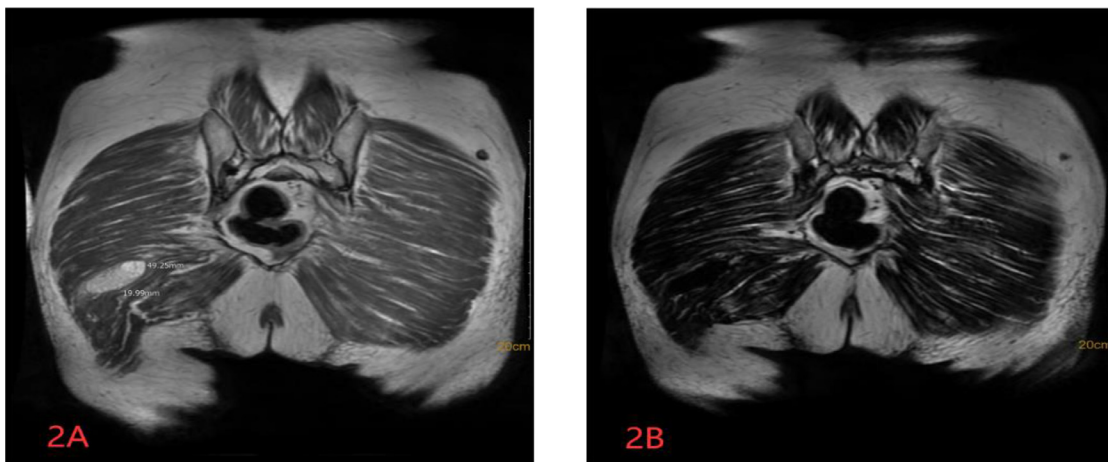


Fig. 2 – MRI revealing a 1.9 cm × 4.9 cm mass in the right gluteus maximus (A, B) in 2019

referred to our hospital with a chief complaint of a gradually increasing mass in his buttock. The magnetic resonance imaging (MRI) was performed and showed a 3.9 cm × 2.0 cm mass in the inferior right gluteus maximus, which presented isointense on T1-weighted images, hyperintense on T2-weighted images, and diffusion weight imaging with clear margins (Figs. 1A and 1B). After accurate evaluation, the patient was operated under general anesthesia. Intraoperatively, the tumor was subsequently exposed around 3 cm × 4 cm × 5 cm and completely excised from margin with careful removal of all residual tumors and attached muscles. Routine postoperative pathological examination confirmed that the final diagnosis was chordoma.

Postoperatively, the patient was followed up at our hospital. In 2019, he experienced discomfort again and had presented with a mass the “size of an apricot” in his right hip. MRI signals of the mass were isointense or hypointense on T1-weighted images and hyperintense on T2-weighted images at the right inner gluteus maximus (Figs. 2A and 2B). Given his medical history and this finding, we suspected that the mass was a chordoma. Evidently, recurrence took place

despite previous extensive resection. The patient refused to undergo operative therapy although all conditions were suitable. The patient was discharged without any special treatment.

In 2020, the patient arrived at our hospital for the fourth time because the size of the gluteal tumor was slowly increasing during the past one year. He reported gentle compressive pain and left hip pain. The enhanced MRI revealed a 5.6 cm × 2.6 cm × 2.8 cm irregular space-occupying lesion with unequal intensification just like “vegetable-sponge” intensity at the right inner gluteus maximus (Figs. 3A and 3B). The giant mass and surroundings were circumscribed and extensively completely resected. The routine postoperative pathological examination confirmed the diagnosis of gluteal chordoma. Immunohistochemical staining showed that the chordomas were positive for cytokeratin, epithelial membrane antigen, S-100, vimentin, CK8/18, CD34, and Ki-67 (10%) but negative for desmin.

The patient was discharged without any complication or adverse event 10 days after surgery. The 10-month follow-up revealed that the patient had not undergone adjuvant treat-

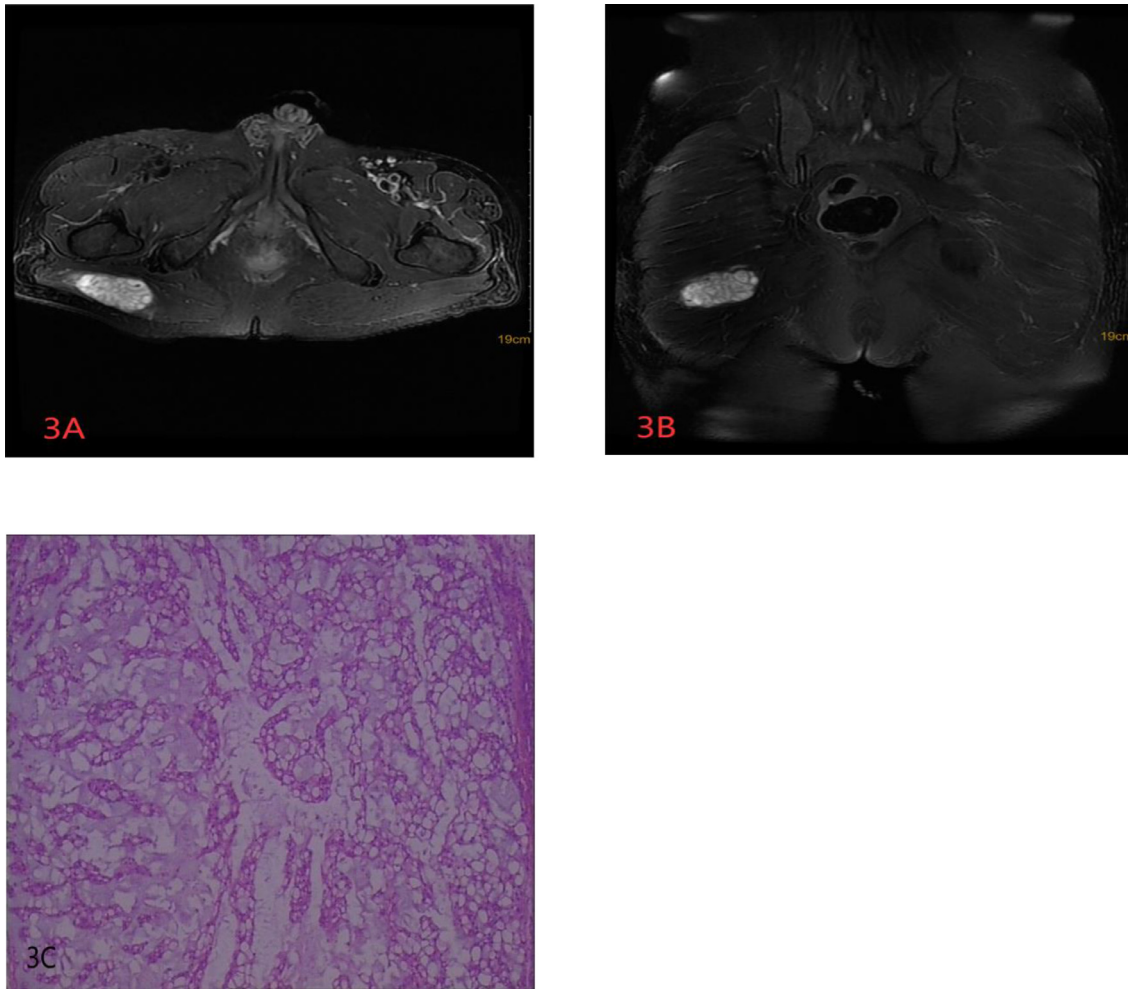


Fig. 3 – Irregular mass with unequal intensification just like “vegetable-sponge” intensity on MRI (A and B). Photomicrographs showing physaliphorous cells within a myxoid stroma observed using H&E staining (C) in 2020

ment and his lower-extremity discomfort and numbness were resolved.

Discussion

Chordoma originating from the remnants of the embryological notochord accounts for 1%–4% of the malignant bone tumor.^[7] The local recurrence rate is 41.3%, and the distant metastasis rate is 34.8%^[8]. In the present case, the metastasis and recurrence of chordoma at the right gluteus maximus following sacrococcygeal region are extraordinarily rare. Muscle metastases are extremely rare because the chemical microenvironment of muscle tissue, like anticancer proteases and cytokines, prevents tumor from the metastasis and mechanical contraction of muscle tissue may stop microemboli from entering the muscle tissue^[9]. We encounter a rare case of chordoma in the gluteus maximus of a 73-year-old man, whose symptoms are gradually extensive mass, lower-extremity discomfort, and numbness. To date, brachyury, a transcription factor of the T-box family, is a sensitive and specific marker

and may serve as an important diagnostic and prognostic biomarker^[10].

The presence of metastases is significantly associated with an increased rate of local recurrence^[8]. Several studies showed that the survival period of patients with metastasis is significantly shorter than that of patients without metastasis^[11]. The distant metastasis is associated with a 3- to 4-fold reduction in patient survival^[12]. Andrew ^[13] found that the CSPG4 expression is associated with metastasis and high mortality because of its involvement in signaling pathways associated with cell proliferation, survival, and migration. Recent studies showed that the tumor marker Ki67 may predict metastatic spread. Specifically, a Ki67 positivity level of higher than 5% is correlated with increased risk of metastatic disease^[9]. In this case, the Ki67 is 10%. Toru Akiyama^[8] showed that recurrence occurs primarily in the surrounding muscle especially the gluteus maximus. Interestingly, this patient is still alive after suffering from metastatic chordoma recurring in the gluteus maximus.

The first choice of treatment of chordomas is surgical resection, and whether postoperative adjuvant radiotherapy is performed should be based on the patients' condition^[14]. In

the present case, the patient declined radiotherapy for personal reasons, explaining why he had metastatic chordoma and recurrence to some extent. Multimodal treatment approaches have significantly altered the therapeutic paradigms for patients with chordoma over recent decades[15]. In patients with inoperable and progressive disease, imatinib has been shown to arrest tumor growth[16]. In 2019, Arish Noor described multiple surgical resections, radiotherapy session, and afatinib therapy of a chordoma from C3 to C6 in a 68-year-old female experiencing neck pain and motor weakness. This female patient remained positively responsive to afatinib treatment.

Conclusion

We encountered an extremely rare metastatic chordoma in the gluteus maximus with muscle recurrence following sacrococcygeal chordoma. In the present case, radiography typically reveals an irregular heterogeneous-intensity mass with clear boundaries in the gluteus. Favorable outcomes remarkably rely on early diagnosis considering the patient's medical history, imaging results, and pathological findings. Hence, long-term observation and further research should be performed to elucidate the nature of chordoma and obtain an understanding of diagnosis and treatment.

Patient Consent

Written informed consent was obtained from the patient we reported.

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