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Case Report

Intra-muscular extraskeletal myxoid chondrosarcoma of the thigh: a case report

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Abstract: Soft tissue sarcomas are mesenchymal tumors that account for about 1% of all malignancies. We retrospectively analyzed a rare case of a painful intra-muscular extraskeletal myxoid chondrosarcoma in the thigh of a 35-year-old man, that had undergone excision. Histological and immunohistochemical analysis of the mass revealed extraskeletal myxoid chondrosarcoma. The patient proceeded to radiotherapy and chemotherapy after curative surgery and had a good outcome.

Keywords: Extraskeletal myxoid chondrosarcoma, intra-muscular, thigh, pain

Introduction

Soft tissue sarcoma is a common malignant tumor derived from mesenchymal tissue. Chondrosarcoma, a malignant tumor of cartilage, occurs most commonly in middle-aged adults between 30 and 60 years old and originates in the extremities and the metaphysis of long bones [1]. Extraskeletal myxoid chondrosarcoma (EMC) is a relatively rare and poorly characterized tumor, that typically arises in the deep soft tissue of the extremities [2]. Here we report a patient with undifferentiated chondrosarcoma in the gluteus maximus muscle.

Case presentation

A 35-year-old male patient presented with a rounded mass on the posterior right thigh that was of 6 months' duration. Grossly the rounded mass was 10 × 10 cm in size, with a hard texture, moderate texture, and above the skin surface. In addition, we examined both the ultrasonography and MRI after hospitalization. Doppler ultrasonography of the mass showed that the neoplastic lesions were located in the subcutaneous adipose tissue with abundant blood flow. On MRI examination a 9.0 × 4.8 cm mass was centered in the lower edge of the right gluteus maximus muscle, which displayed a slightly longer T1 (**Figure 1A**) and longer T2 signal intensity (**Figure 1B**). The report described a solid space-occupying cystic lesion on the lower border of the right gluteus maximus muscle and was highly suspicious for tumor. The serological tests showed an increased neuron-specific enolase of 24.10 ng/ml.

Based on the results of these examinations, we performed tumor resection from the right thigh root. The operative report is as follows: the patient was placed in the prone position after spinal anesthesia and standard disinfection and draping were carried out. We made a 15 cm skin incision over the tumor on the root of the right thigh. After separating the subcutaneous tissue, we exposed an intermuscular mass, which displayed a firm texture, grayish white and multinodular with a clear boundary. Grossly, the tumor had a very glossy cut surface and a complete capsule with translucent tumor tissue. We dissociated the muscle surrounding the tumor and found that the surrounding muscle was compressed by the tumor. After the muscle tissue around the tumor was separated

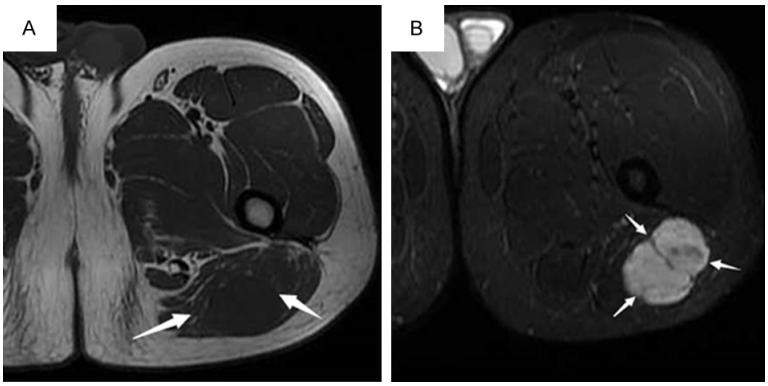

Figure 1. Magnetic resonance imaging (MRI) reveals the lesion of a lobular mass. A. Coronal T1W SE image shows a homogeneous intermediate SI mass (arrows) with muscle erosion (arrowhead). B. Axial fat-suppressed T2W FSE image shows the hyperintense mass (arrows) lying deep in the muscle (arrowhead).

and cut off, the tumor was removed entirely. The tumor sample was subjected to pathologic examination.

Grossly the cut surface of the tumor revealed a round shape, with elastic firm, slightly lobulated nodules, grayish-white, jelly-like, with mucoid material in some areas, without obvious necrosis or cystic degeneration (**Figure 2A**). Microscopic examination showed round, oval-shaped, or spindle cells that were arranged in cord-like structures (**Figure 2B**). H&E staining showed that the cytoplasm was eosinophilic to pale, the nuclei were round-shaped or oval-shaped and mitosis was rare (**Figure 2C**, **2D**). The thin fibrous septa were abundant in the myxoid matrix and showed a lobulated shape. Although vessels were rare in the mass, hemorrhage, necrosis, or cystic degeneration were found. Focal necrosis was observed in some sections.

The immunohistochemical (IHC) staining showed that the tumor cells were positive for these markers: Vimentin, INI1, S-100 (+) (**Figure 2E**), local expression of EMA, and CD34. Tumor cells were negative for these markers: SMA, CD117, Syn, ERG, and p63. The Ki-67 expression level of tumor cells was about 5% (**Figure 2F**). The myxoid stroma was positive for AB-PAS staining. Pathologic diagnosis was extraosseous myxoid chondrosarcoma (EMC).

After surgery, the patient had radiotherapy in combination with chemotherapy and achieved a satisfactory therapeutic effect. At the two-year follow-up, the patient did not present any clinical signs of EMC, and no mass was found on re-examination. MRI re-examination was confirmatory.

Discussion

Extraskeletal chondrosarcoma, accounting for less than 3% of all soft tissue sarcomas (STS), is a rare mesenchymal malignant tumor with a multidirectional differentiation capacity [3]. About 80% of extraskeletal chondrosarcoma usually occurs in the deep tissues of the proximal limbs and limb girdles. The neoplasm is categorized as an indeterminate malignant neoplasm according to the 2016 WHO classification of soft tissue sarcoma [4].

Extraskeletal chondrosarcomas are typically divided into myxoid, mesenchymal, and well-differentiated types based on histologic criteria [3]. The patient's clinical characteristics include subcutaneous mass, pain, bleeding, and dyskinesia. Although the etiology is not clear, some studies reported that the neoplasm is related to genetic alterations or abnormalities of soft tissue [5]. Myxoid chondrosarcoma is a slow-growing tumor and can be removed by complete surgical resection. Mesenchymal chondrosarcoma shows a high incidence of lung and liver metastases and can be treated with a combination of chemotherapy and radiotherapy after surgical removal. A well-differentiated chondrosarcoma is extremely rare.

The main differential diagnosis includes neoplasms that produce mucin-rich carcinoma, such as chordoma, chondrosarcoma, osteoblastoma, and myoepithelial carcinoma. Our reported case is a typical extraosseous myxoid chondrosarcoma, and the noticeable symptoms included pain in the upper thigh.

Extraskeletal myxoid chondrosarcoma (EMC) is an aggressive malignant soft-tissue sarcoma with prominent myxoid stroma, that mostly occurs in male patients aged 30-60 years [6]. The median age is 52 years and the male-to-female ratio is 2:1. EMC usually develops in the

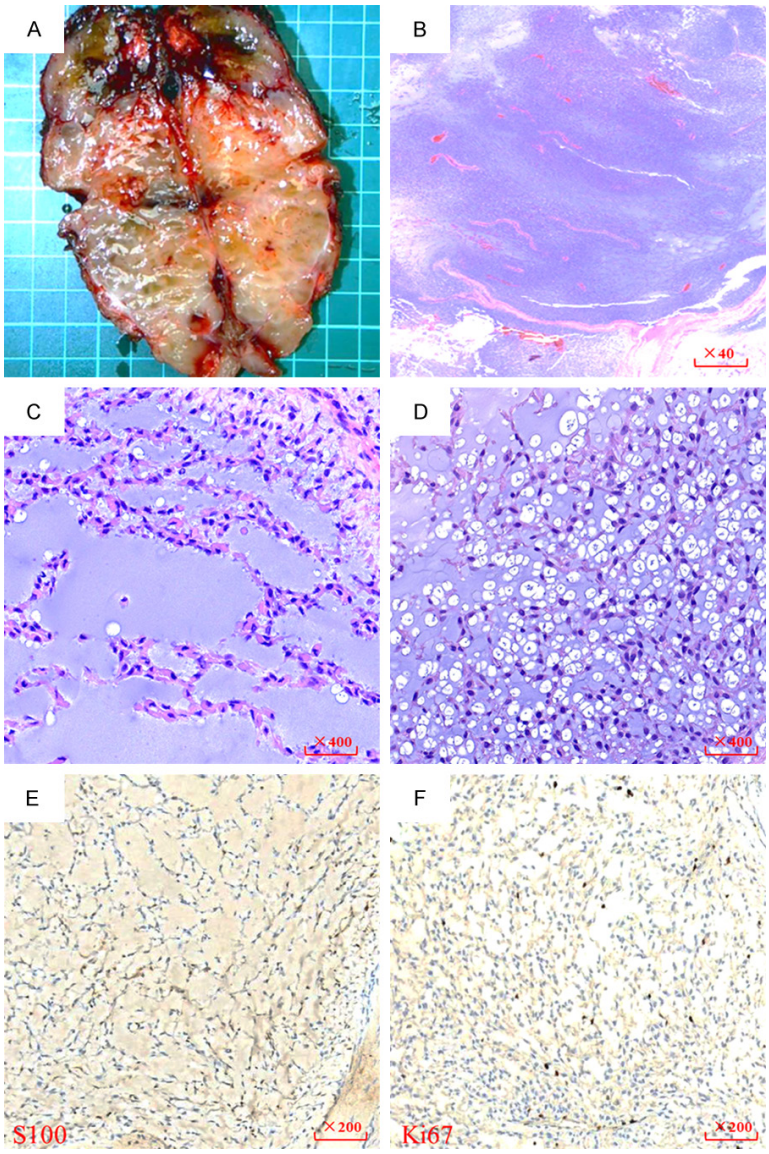

Figure 2. Morphology of extraskeletal myxoid chondrosarcoma. (A) Gross morphology shows the cut section of the surgical specimen. (B) Histopathologic morphology reveals a nodular pattern with areas of primitive round cells (H&E, × 40). (C) There are spindle-shaped cells and myxoid stroma. (H&E, × 400). (D) Chondroid tissue (H&E, × 400). Immunohistochemical staining for S-100 (E) and Ki67 (F) (× 200).

deep soft tissue of the proximal extremities and limb girdles, with the thigh being the most common primary site [7]. Other less common sites are the meninges, orbit, and brain. The average diameter of EMC is 7 cm [8]. The sarcoma is slow-growing, with slight local swelling and mild pain. The patient's findings were consistent with previous descriptions. The location and size of the tumor were consistent with the described clinical symptoms. However, the patient age was relatively young, at 35 years old.

Clinical treatment: Patients with chronic severe pain are given analgesics to reduce pain. As chemotherapy alone is ineffective, many patients generally require adjuvant chemotherapy and radiotherapy followed by radical surgical resection [9]. Patients with ECM exhibited high survival rates of approximately 80% at 5 years and 60% at 10 years. A few patients still developed local recurrence at a follow-up of 6-84 months [10]. Our patient got surgical resection and postoperative radiotherapy and chemotherapy. The patient had a good outcome without recurrence at 2 years of follow-up.

Acknowledgements

A signed informed consent was obtained from the patient.

Disclosure of conflict of interest

None.

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