CASE REPORT

The CT and MRI Findings of Sacral Mesenchymal Chondrosarcoma: A Case Report

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Mesenchymal chondrosarcoma is a rare malignant tumor. It may exhibit as a skeletal or extraskeletal tumor and usually contains calcified deposits. A lesion arising from sacrum is extremely rare. We present a 17-year-old male patient with clinical manifestations of lower back pain and sciatica. Computed tomography and magnetic resonance images of the patient depicted a poorly demarcated sacral tumor with a large extrasosseous soft tissue component, but without calcification. Multiple small foci of lung, bone, and lymph node metastases were also found on staging CT images. CT guided biopsy and lumbosacral laminectomy (L5-S3) with partial removal of extradural mass for decompression were performed. The histopathological diagnosis was mesenchymal chondrosarcoma. However, the tumor had poor response to adjuvant chemotherapy and radiation therapy during an 8-month follow-up.

Key words: Mesenchymal Chondrosarcoma; Sacrum

Mesenchymal chondrosarcoma (MCS), first described by Lichtenstein and Bernstein in 1959 [1], is a rare form of chondrosarcoma. It may originate from bone or soft tissue. Only few cases of MCS from the sacrum have been reported in literatures [2,3,4].

CASE REPORT

A previously healthy 17 y/o boy suffered from progressive lower back pain with radiation to left lower extremity for one month. Progressive left leg weakness with difficulty in urination, defecation, and erection were also noted. Then he was admitted to our hospital. The initial neurological examination revealed decreased muscle power over distal left lower extremity, decreased pinprick sensation at the dermatome of left S1-S3, decreased left ankle jerk, and negative for Babinski' reflex. The laboratory data was grossly normal.

Plain film showed an osteolytic lesion around the superior margin of left sacrum (Fig. 1). Computed tomography (CT) demonstrated a poorly demarcated pelvis mass with bone destruction of the sacrum and the 5th lumbar vertebra. Paraspinal muscle infiltration and epidural extension were also found, but no calcification was detected. The lesion was isodense to normal muscle on unenhanced CT and had heterogeneous enhancement following contrast injection (Fig. 2). Magnetic resonance imaging (MRI) for lumbosacral spine also depicted the lesion, which was isointense to muscle on conventional spin echo T1-weighted images (CSE T1WI, TR/TE=752/15 ms) and heterogeneously hyperintense on gradient echo.
T2-weighted images (GRE T2WI, TR/TE=808/18 ms, FA=20°), and had inhomogeneous enhancement (Fig. 3a,b,c). Small foci of lung, paraaortic lymph nodes and lumbar spine metastases were also noted on staging CT images. Multiple bony metastases were suspected by bone scan. The differential diagnosis on the basis of radiological findings and clinical manifestations include lymphoma, primitive neuroectodermal tumor, small round cell sarcoma, metastases or MCS.

He underwent CT guided biopsy and lumbosacral laminectomy (L5-S3) with partial removal of extradural mass for decompression. The microscopic features showed biphasic pattern with primitive hyperchromatic tumor cells arranged around a pericytomatous vasculature and the chondromyxoid background. Minimal foci of calcification were found (Fig.4). The histochemical stain revealed that the chondromyxoid tissue was positive for mucicarmine and alcian blue at pH 1.0, and the primitive tumor cells were positive for MIC-2, equivocal for synaptophysin, but negative for chromogranin, LCA, cytokeratin and desmin, PAS and S-100. Although no typical mature chondrocyte was detected, the microscopic and histochemical study supported that the mesenchymal tumor cells had a tendency toward chondrocytic differentiation. The histopathologic diagnosis is a mesenchymal chondrosarcoma, which had been confirmed by consultant pathologist, Sharon W. Weiss.

Because of local invasion and multiple metastases, adjuvant radiotherapy and chemotherapy were attempted. However, after an 8-month period of follow-up and treatment, his clinical status became downhill.

**DISCUSSION**

Mesenchymal chondrosarcoma is a rare variant of chondrosarcoma and may arise from bone or soft tissue. It tends to occur in second and third decade (60-70%) without sex predilection [4,5,6]. Osseous MCS preferentially locates in the jaw, maxilla, femur, and rib [4,6]. In contrast, extraskeletal MCS mostly involve craniospinal meninges and lower extremity [4,6]. About 10% of MCS involve the pelvic girdle, but MCS from sacrum is rare [2,3,4,5].

The typical skeletal MCS is an osteolytic tumor, with cortical destruction, extraosseous extension, and stippled or amorphous calcification. Regardless of skeletal or extraskeletal MCS, MR imaging often discloses an iso- or hypointense tumor on T1WI, which is heterogeneously hyperintense on T2WI. Different
degree of tumor enhancement depends on the cellular and chondromyxoid contents [7]. The images of our patient demonstrate similar features, except for radiographically detectable calcification. However, absence of calcification cannot exclude the possibility of MCS. According to the literatures, the incidence of calcification ranges from 67% to 100% [4,5].

The radiographic findings of MCS are nonspecific. Definite diagnosis relies on histopathological examination, which discloses biphasic components of small undifferentiated cells and cartilage islets around a pericytomatos vasculature and the chondromyxoid background. The differential diagnosis includes other small round cell sarcoma, PNET, lymphoma and metastasis [3,8]. Although no typical mature chondrocyte was detected in our patient, the microscopic and histochemical study supported that the mesenchymal tumor cells have a tendency to chondrocytic differentiation. It might be the reason that only minimal foci of calcification was detected in microscope.

Complete surgical resection is the most
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effective treatment. But radical surgery cannot be performed when extensive local invasion or distant metastases is accompanied. Adjuvant radiotherapy and chemotherapy may be arranged for unresectable and metastatic tumor. Effect of radiotherapy and chemotherapy is still controversial. Some reports show response in unresectable tumor [5,9], but others show no significant effect in prolonging survival time [4]. Anyway, the prognosis of MCS remains poor due to high metastatic or recurrent rate. According to the report of Nakashima et al., 73.9% of patients die of the disease 6 months to 23 years after diagnosis, with average of 6.7 years. Local recurrence or distant metastases, most commonly in bone and lung, may be encountered even 20 years after diagnosis [4].

In conclusion, the MCS is a well-established pathologic entity. Lesions from sacrum are extremely rare. Lack of radiographically detectable calcification cannot exclude the diagnosis. Although no enough data available, the radiographic or pathologic findings of MCS from sacrum have no difference from that of the other site. The prognosis may be poor due to delayed diagnosis and difficulty in radical surgical approach.

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REFERENCES

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骨間質軟骨肉瘤在電腦斷層和磁振造影的影像表現—病例報告

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間質軟骨肉瘤一種罕見的軟骨悪性腫瘤，而發生於骨的間質軟骨肉瘤更是罕見。我們報告一個十七歲的年輕男性病例，因為逐漸惡化的下背痛和坐骨神經痛來求診，電腦斷層和磁振造影呈現一個間質腫瘤，經由電腦斷層導引下組織切片及開刀後，確立最後病理診斷為間質軟骨肉瘤。然而這個病人的影像並未呈現典型的間質軟骨肉瘤鈣化現象。我們將對於他的臨床表現和影像做一個簡單的討論。

關鍵詞：間質軟骨肉瘤，骨