

Chondrosarcoma of the dorsal spine – A rare case

ABSTRACT

Chondrosarcomas of the spine are rare tumors and represent <10% of all chondrosarcomas. In the spine, they may arise from vertebral bodies or posterior elements. They may occur in patients ranging from 13 to 78 years of age. Here, we present a case of a 25-year-old female who presented with complaints of lower backache, stiffness in both lower limbs, and tingling sensation in the right lower limb. On examination, both power and sensations were decreased below waist. Magnetic resonance imaging spine revealed an extradural lesion at D5 vertebral body level with severe cord compression. We received the mass in multiple fragments which were grayish-white and firm to hard in consistency. Microscopically, a chondroid tumor was seen with cells arranged in lobules in abundant myxoid matrix. The neoplastic chondrocytes were large in size and had bizarre hyperchromatic nuclei. Few binucleate and multinucleate forms were also seen along with occasional atypical mitoses. There was permeation and destruction of the host bone, and the tumor was seen invading the marrow spaces. Few foci showed high cellularity. No osteoid formation was seen by the tumor. The tumor was diagnosed as Chondrosarcoma – Grade II.

Keywords: Chondroblastic osteosarcoma, chondroma, chondrosarcoma, chordoma, spine

INTRODUCTION

Chondrosarcoma is a malignant mesenchymal tumor with cartilaginous differentiation.^[1] According to the WHO classification, Grade I chondrosarcoma or atypical cartilaginous tumor is grouped under locally aggressive (intermediate) cartilaginous tumors, whereas Grade II and Grade III chondrosarcomas along with Clear cell, mesenchymal, and dedifferentiated subtypes are classified as malignant chondrogenic tumors.^[2] Chondrosarcoma is the third most common primary malignant bone tumor; however, spinal chondrosarcomas are rare and represent <10% of all chondrosarcomas.^[3] Incidence of spinal chondrosarcomas is estimated to be between 2% and 12% according to various series.^[4] Here, we report a rare case of chondrosarcoma of thoracic spine in a young patient.

CASE REPORT

A 25-year-old female with no history of any chronic medical illness, presented with complaints of lower backache, stiffness and weakness in both lower limbs, and tingling sensation in the right lower limb for 1½ months. On examination, the power was 3/5 in both lower limbs proximally and 2/5 and

1/5 distally in the right and left lower limbs, respectively. Sensations were decreased below the waist.

Magnetic resonance imaging of the spine revealed a heterogeneously enhancing 25 mm × 19 mm extradural lesion at D5 vertebral body level, causing displacement of the cord to the right and severe cord compression [Figure 1].

Intraoperatively, D5 level extradural soft-tissue lesion was seen, which was granular and appeared to have a rich vascularity.

Mass was received in neuropathology department in multiple small bits and fragments aggregating to

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2.5 cm × 2 cm × 0.5 cm. Bits were grayish-white in color and firm in consistency. Few bits appeared bony hard.

On microscopic examination, multiple biopsy bits showed a chondrogenic tumor with cells arranged in lobules in abundant myxoid matrix. The neoplastic chondrocytes were large in size and had bizarre hyperchromatic nuclei. Few binucleate and multinucleate forms were also seen along with occasional atypical mitoses. There was permeation and destruction of the host bone, and the tumor was seen invading the marrow spaces. Few foci showed high cellularity. No osteoid production was seen by the tumor. The tumor was diagnosed as Chondrosarcoma – Grade II [Figures 2-4].

DISCUSSION

Spine is a rare location for chondrosarcomas where it can present as a primary tumor or may develop as a secondary tumor in an already preexisting benign cartilaginous lesion.^[1] Long bones, ribs, shoulder, and pelvic girdles are common locations for chondrosarcoma,^[5,6] and only 6%–10% are known to occur in the spine.^[7] Epidemiological data indicate a wide range of age from 13 to 78 years with a mean age of 33 years, and there is almost equal distribution of cases among both sexes.^[8,9] Huvos and Marcove reported that spinal origin in chondrosarcoma was more common in patients younger

than 21 years of age than in adults.^[4] The present case was a 25-year-old female.

Most of the spinal chondrosarcomas occur in the thoracic spine followed by cervical region and are relatively rare in lumbar vertebra. They can arise from the body and posterior elements of the vertebral column, but are more frequently reported to arise from the latter.^[10-12] In our case, tumor was seen arising from the vertebral body of 5th thoracic spine vertebra.

Some of the commonly reported symptoms of spinal chondrosarcomas include pain, swelling, sensory and motor deficits resulting from spinal cord compression, and pathological fractures.^[12-14] The present case had a severe cord compression, resulting in both sensory and motor deficits.

The chondrogenic tumors of bone are now classified into benign, intermediate (locally aggressive), intermediate (rarely metastasizing), and malignant grades. Chondrosarcoma Grade I, also called as atypical cartilaginous tumor, is classified as “Intermediate” (locally aggressive) tumor, whereas Grade II and III chondrosarcomas along with clear cell, mesenchymal, and dedifferentiated subtypes are classified as “Malignant.”^[2,10] Grade I tumors have low cellularity and lack pleomorphism; they have a hyaline cartilage matrix, and they metastasize very rarely. Grade III chondrosarcomas have a very high cellularity with pleomorphism and mitotic figures. They often have muco-myxoid matrix and metastases

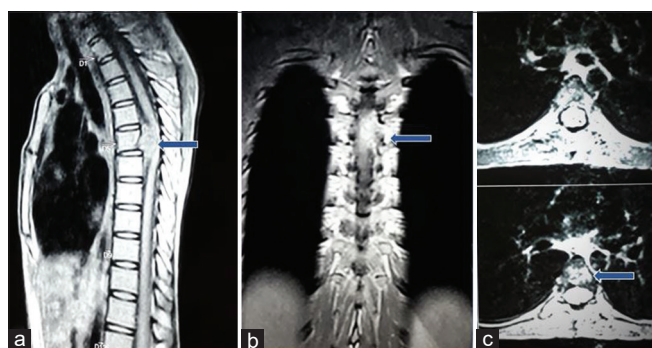


Figure 1: (a) T1 weighted sagittal image of thoracic spine, (b) Post contrast T1 weighted coronal image of thoracic spine and (c) Post contrast T1 weighted axial image at D5 level

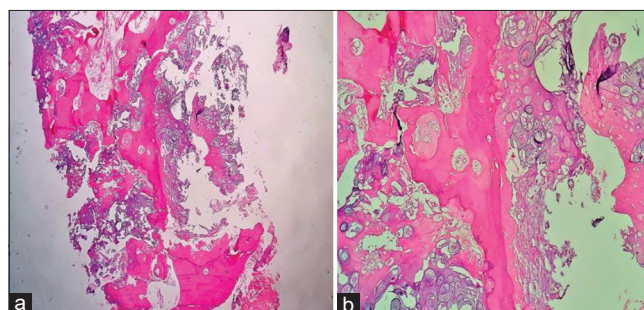


Figure 2: (a) (H and E, ×4) and (b) (H and E, ×10) A cartilaginous tumor showing vague lobules of neoplastic chondrocytes with permeation of the host bone and infiltration of the marrow spaces

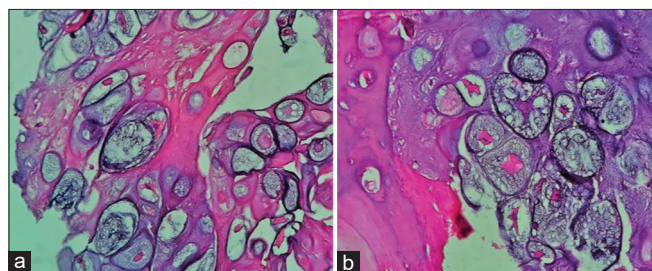


Figure 3: (a) (H and E, ×40) and (b) (H and E, ×40) The neoplastic chondrocytes were large in size and had bizarre hyperchromatic nuclei. There was abundant myxoid matrix

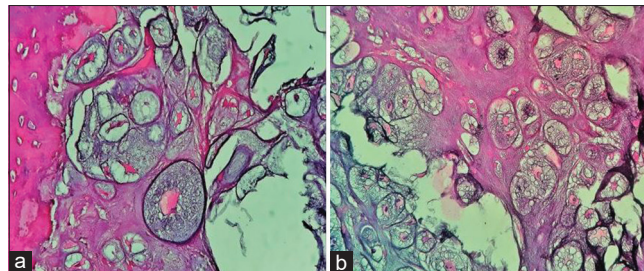


Figure 4: (a) (H and E, ×40) and (b) (H and E, ×40) Few binucleate and multinucleate forms were seen along with foci of higher cellularity. Occasional atypical mitoses were seen elsewhere

occur in 70% of patients. Grade II chondrosarcomas have characteristics of both Grade I and Grade III tumors. Variant subtypes of chondrosarcoma such as clear cell, mesenchymal, and dedifferentiated are extremely rare in the spine.^[10,15] Strike and McCarthy in their case series of 16 spinal chondrosarcomas encountered tumors of all three grades. However, there was only one Grade III Chondrosarcoma, and majority were Grade II followed by Grade I. They also concluded that despite the fact that majority of the spinal chondrosarcomas are Grade I and Grade II tumors, many are lethal due to higher risk of pulmonary metastasis. Incomplete resection is a major contributing factor.^[3] In our case, we encountered few foci of high cellularity with increased pleomorphism and mitotic activity. There was also infiltration and destruction of the host bone. The overall features were intermediate between Grade I and Grade III; hence, it was classified as Chondrosarcoma Grade II. We encountered abundant myxoid stroma formed by the tumor. Myxoid change is related to malignancy in cartilaginous tumors, or it may indicate degenerative change. Degenerative myxoid change is characterized by the presence of myxoid areas without significant cellularity, whereas the myxoid change associated with malignancy is characterized by loss of lacunar pattern with pleomorphic spindle or stellate cells floating in a myxoid stroma.^[15] In our case, stellate cells were seen floating in the myxoid stroma associated with foci of higher cellularity.

Three main differential diagnoses have to be considered in any case of the spinal chondrosarcoma. Benign cartilaginous tumor enchondroma is extremely rare in spine, but it can closely mimic chondrosarcoma, especially low grade. It has the same lobular architecture and can have binucleate cells and necrosis. However, radiographically, it is benign and bone invasion, and destruction is usually not observed.^[15] Diagnostic difficulty can arise in cases of Grade I chondrosarcomas; however, in the present case, tumor showed areas of high cellularity with increased mitotic activity. Furthermore, bone invasion and destruction were observed, so this differential was not considered. Similarly, chordomas, which are common in skull base and sacral region, can closely mimic chondrosarcomas, especially the chondroid chordomas. However, chordomas of the thoracic spine are extremely rare.^[16] Many times, immunohistochemistry is needed to differentiate between the two.^[3] In the present case, however, clear cut cartilaginous differentiation was observed, and no arrangement of cells in cords or physaliphorous cells were seen, so no immunohistochemistry was needed to rule out a chordoma. Chondroblastic osteosarcomas can be extremely difficult to diagnose on small biopsies, as multiple sections need to be examined to detect the presence of neoplastic osteoid among the cartilage.^[3,15] In the present case, the entire

specimen received was processed and extensively examined for the presence of osteoid. After confirming the absence of osteoid, the tumor was diagnosed as Chondrosarcoma.

CONCLUSION

Spinal chondrosarcomas are rare tumors, which can arise from the body of the vertebra or from the posterior elements of the vertebral column. Although majority are Grade I and Grade II tumors, Grade III tumors can rarely be encountered. Chondroblastic osteosarcoma needs to be ruled out in all the cases along with benign chondrogenic tumors such as chondroma and also midline chordomas chordomas.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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