



MESENCHYMAL CHONDROSARCOMA OF THE SPINE, A GIANT RETROPERITONEAL MASS: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Spinal chondrosarcomas are rare lesions of spine and should be completely resected and resistant to adjuvant chemotherapy and radiotherapy. Partial removal of the tumor with the combination of adjuvant radiotherapy may be preferred for palliation of symptoms for advanced stage tumors. Here in, we present a 48-year-old male patient having a giant retroperitoneal mesenchymal chondrosarcoma originated spinal column.

Malignant lesions on the spine are challenging cases for both orthopedic surgeons and neurosurgeons. After years of experience, these tumors can be successfully treated. These operations, which have catastrophic complications such as paraplegia or tetraplegia, infection and sepsis, need to be performed in experienced centers. Under the light of this work and the literature, the idea was obtained which is it is necessary to make good planning before the first surgery in the primary chondrosarcomas of the spine and to protect the neural structures from being easily sacrificed during mass excision and to preserve the enblock total vertebra excision in order to prevent recurrence, prolong the life expectancy and improve the comfort of life.

Key words: Mesenchymal chondrosarcoma, paraplegia, primary malign spinal tumors

Level of evidence: Case report, Level IV

INTRODUCTION

Chondrosarcoma is an uncommon malignancy, which arises from a cartilaginous origin that accounts for 10 % of primary bone tumors ⁽¹⁵⁾. Lichtenstein and Bernstein first identified them in 1959. Chondrosarcomas are classified into three subgroups, which are myxoid, mesenchymal and well differentiated. Mesenchymal chondrosarcomas are infrequent, aggressive and has extremely poor prognosis with rapid progression compared with the other two subtypes. Extra skeletal localizations are infrequent. Orbit, cranial and spinal meningeal sheets, lower limbs are where the most frequently observed ⁽⁷⁾.

En-bloc resection in the surgery is the preferred form of treatment. Palliative resection with or without chemotherapy and radiotherapy may reduce symptoms, especially pain when resection is not possible or in case of distant metastasis, but life expectancy is extremely short. In this paper, we present a 48-year-old male

patient having a giant retroperitoneal mesenchymal chondrosarcoma.

CASE REPORT

A 45 year-old male patient admitted to clinic with complaint of abdominal tenderness, distension and pain. In his history, he underwent tibial surgery 8 years ago with the pathological diagnosis of osteosarcoma. After 6 months, cause of metastatic lung cancer, he had been operated four time for partial lung excision. Since 6 years he had well health and he had started to the work. One year ago, patient had complains of severe back pain and weakness in the legs. After physical and radiological examination, he diagnosed to have a spinal tumor in lumbar region. He had an unsuccessful surgery and tumor mass could not be removed also he had paraplegia after surgery. Two months after the operation, the patient admitted the hospital with severe fever. The patient was diagnosed as sepsis and who needs intensive care is referred to our hospital. In

the patient's examinations, leukocytes are 13000/mm³(4600-10200/mm³), c-reactive protein is 15.2 gr/dL(0-0.5mg/dL). Enterococcus faecalis is found in the culture taken from the puss in the surgical area.

The patient has taken to the operation. In the prone position, old incision was used. After debridement, posterior instrumentation was removed. Laminectomy area was widened and medulla spinalis damaged was noticed. Than posterolateraly, the tumoral mass was excised totally and the titanium cage with the bone cement was put in the vertebrectomy side posterolateraly. After the operation, he is recovered from sepsis. The patient is discharged on the 21st day after surgery.

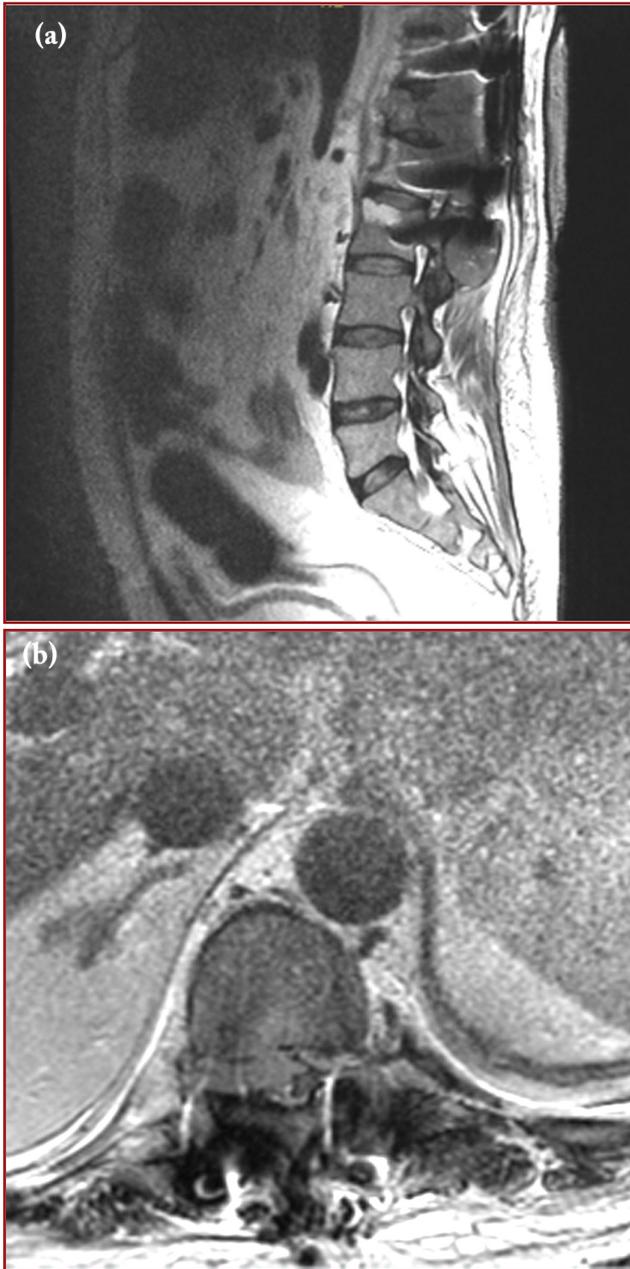


Figure-1. a) Sagittal and **b)** axial MR imaging of the patient with sepsis. Abscess formation was seen in the paravertebral area. Complete damage of spinal cord has been noticed.

Six months after the last operation, the patient admitted to clinic with a complaint of abdominal tenderness, distension and pain. In computed tomography (CT), two retroperitoneal localized, calcified, necrotic masses were found which are 20 cm and 35 cm in diameter. The patient undergone debulking surgery. Both masses are removed. During surgery, it was seen that, masses originated from the lumbar spine. Patient was followed up in the intensive care unit for 5 days and discharged on the 14th day. The result of the pathologic specimen was malign mesenchymal chondrosarcoma.



Figure-2. Two giant tumor specimens after the tumor excisions.

DISCUSSION

Chondrosarcomas are cartilaginous matrix synthesizing tumors and most of the chondrosarcomas arises from bones or cartilages. Chondrosarcoma is the third common malignant bone tumor following osteosarcoma and Ewing's sarcoma⁽⁴⁾.

Chondrosarcomas are commonly located in the thoracic vertebrae and dominant in male gender. Cervical and lumbar vertebrae are second and third frequent locations⁽¹³⁾. Extraskeletal localizations are rare. Orbit, meningeal sheets

of central nervous system and lower limbs are frequent Extraskelatal locations⁽⁷⁾. Malignant tumor originated from lumbar spine in our patient. After completing the skeletal maturation, osteochondromas and enchondromas should be suspected of chondrosarcoma transformation if they grow or become painful. Chondrosarcoma of the osteochondromas is thickened by cartilage capillary lesions. Spread of lesion in neighboring vertebral segments taking into the disc is seen in 35 % of patients⁽¹²⁾.

Well-differentiated and myxoid types grow slowly but mesenchymal chondrosarcomas are more aggressive and tend to make distant metastases.

The type that holds the nervous system of extraskelatal mesenchymal chondrosarcomas is more frequent and most often 3rd and 4th decades. Non-skeletal mesenchymal chondrosarcomas involving soft tissues are seen over 40 years of age⁽¹⁾.

Conventional radiography has no impact on the diagnosis of mesenchymal chondrosarcomas but they mostly appear as destructive lesions in the spine or as a highly opaque calcified paraspinal soft tissue mass in computed tomography or magnetic resonance imaging. Peripheral and focal low attenuation areas are seen in contrast-enhanced computed tomography which are possibly present necrosis⁽¹⁴⁾. In magnetic resonance imaging of mesenchymal chondrosarcomas, intratumoral calcified and non-calcified components demonstrate low and high intensity on T2W1 images. This view of images are named as "salt and papper" sign⁽⁶⁾. Our patient did not have MRI but in CT images two calcified, central necrotic huge masses were found.

As in other malignant spinal tumors, it is difficult to determine the surgical resection boundaries due to the difficulty of local anatomy and the proximity of vital structures in spinal chondrosarcomas^(2,12). Whether vertebra localized or retroperitoneal localized, complete resection of the mesenchymal chondrosarcoma is of critical importance for prolonged survival. When complete resection is performed, 25% cure is obtained. The cases where the marginal resection can not be performed and the relapsed cases result in 74% fatality⁽²⁾. Complete resection with disease free wide margins should be accompanied by a histological examination⁽⁸⁾. For en-bloc resection of spinal chondrosarcoma, a multidisciplinary surgical team is needed including orthopedic, neuro and plastic surgeons⁽¹⁵⁾. Both radiotherapy and chemotherapy are used for adjuvant treatment but positive effect of these therapies on prolonged survival or local recurrence are limited. Better outcome in post operative proton-beam therapy is shown, especially for spinal lesions⁽⁵⁾. Despite all treatment modalities, 5-year survival is 54 %, 10-year survival is 27 %⁽¹¹⁾. The case presented in this study was applied to a brain surgery clinic at another center due to pathological fracture and severe pain at T12-L1 vertebral levels, where marginal resection and posterior instrumentation were performed. This situation has increased the recurrence rate as mentioned above. When the patient was operated on because of paravertebral abscess, the recurrence of rapidly growing tumor mass was noticed.

A titanium cage filled with internal cement was placed to prevent implant failure in the absence of anterior support.

In primary spine malignant tumors, neurological deficits occur in up to 20 % of tumors due to pressure or malignant invasion or due to arterial compression, occlusion and ischemia⁽¹²⁾. In the presented case, postoperative paraplegia development shows that neural structures are injured during mass excision. If complete resection of the tumor is not possible, partial removal followed by radiotherapy may provide palliation in symptoms such as pain or neurological deficiency⁽¹⁶⁾.

Negative prognostic factors are high histological grade, advanced age and primary surgery out of a referral center⁽³⁾. In our patient's history, there were an unsuccessful surgical procedure causing paraplegia and sepsis. After referred to an experienced center in spinal surgery, he had prolonged treatment in intensive care unit for treatment of sepsis and two more operations to correct the complications of the first two operations, and finally the removal of two retroperitoneal masses.

The surgical planning the patient with malign mesenchymal chondrosarcoma is very imported and need wide experience. Instrumentation failure is inevitable result, if the anterior strut grafting or anterior cage or anterior instrumentation would not use. Infection is worst complication after the surgery of spinal tumors⁽¹⁵⁾. Infection rates are very high for the patient's immune system is reduced with chemotherapeutic drugs and the patient is generally in poor condition. The presenting case was also referred to our hospital because of sepsis. Infection with a broad abscess is linked to the surface infection of the implants. After removal of implants, abscess drainage and debridement, the infection could be controlled and the patient could be discharged from the intensive care unit⁽⁹⁻¹⁰⁾.

Malignant lesions on the spine are challenging cases for both orthopedic surgeons and neurosurgeons. After years of experience, these tumors can be successfully treated. These operations, which have catastrophic complications such as paraplegia or tetraplegia, infection and sepsis, need to be performed in experienced centers. Under the light of this work and the literature, the idea was obtained which is it is necessary to make good planning before the first surgery in the primary chondrosarcomas of the spine and to protect the neural structures from being easily sacrificed during mass excision and to preserve the enblock total vertebra excision in order to prevent recurrence, prolong the life expectancy and improve the comfort of life.

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