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Rare Primary Spinal Mesenchymal Chondrosarcoma: A Case Report

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ABSTRACT

Primary extradural mesenchymal chondrosarcoma (MCS) is a very rare intraspinal tumor. Proper clinical evaluation with magnetic resonance imaging (MRI) followed by biopsy of the mass can confirm the diagnosis. Since MCS has a high tendency of recurrence and metastasis, the prognosis is guarded. We report a 52 years old male with primary extradural MCS in the lumbosacral region (L5-S3). Subtotal excision and biopsy were performed which confirmed the diagnosis. The patient received post excisional radiotherapy but expired after four months. Though rare, this tumor should be kept in differentials which helps in early diagnosis and thus can be treated with radical excision of the mass along with postoperative radiotherapy.

Keywords

Chondrosarcoma, extradural, mesenchymal, radiation, spine

INTRODUCTION

Chondrosarcomas are a heterogeneous group of tumors with characteristic ability of cartilage formation. Chondrosarcoma, though the third most common primary malignant bone tumor, rarely presents as a primary tumor of the spine.[1] Mesenchymal Chondrosarcoma (MCS) is a rare malignant variant of chondrosarcoma. Majority of MCS originate from bone but a third of them are detected in extra-skeletal sites which also includes the brain, meninges, and occasionally intraspinal region. Primary mesenchymal chondrosarcoma arising from meninges is extremely rare.[2] We report a case of extradural primary mesenchymal chondrosarcoma in the lumbosacral region arising from the meninges and the results after debulking surgery and postoperative adjuvant radiotherapy.

CASE PRESENTATION

A 52 years old gentleman presented to the spine outpatient clinic with a complaint of low back pain radiating to the left lower limb for duration of four months. The dull aching pain gradually progressed to incapacitating present-day pain over a period of two months. There was no history of trauma and constitutional symptoms suggestive of infection. The spine examination revealed tenderness in the lumbosacral region. Except for absent bilateral ankle reflex, the rest of the neurological examination was normal. The lumbosacral radiographs and routine blood investigations were normal. MRI of the lumbosacral spine revealed extradural mass extending from the body of L5 vertebrae to S3 vertebrae and strongly enhancing after administration of intravenous gadolinium (Figure 1). An excisional

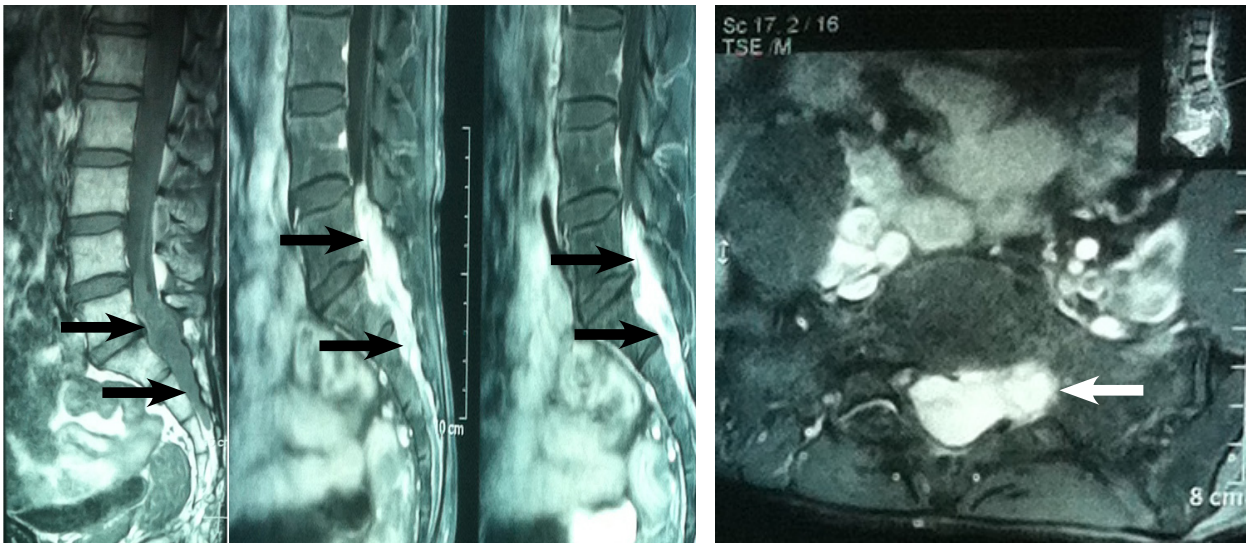

Fig 1. Lumbosacral MRI showing intraspinal mass extending from L5 to S3. Saggital images where the mass appears hypointense on T1 (left) and hyperintense on T2 (middle) with strong enhancement on gadolinium contrast (right). Axial image with gadolinium contrast showing foraminal extension (extreme right).

biopsy was thus planned. The lumbosacral spine was approached by a standard posterior approach. L4 to S3 spinal canal decompression was done by bilateral laminectomy using spinoplasty technique. An extradural grayish-white fluffy mass extending from L5 to S3 was seen. The mass was extensively blended with duramater and extending into the neural foramina of respective nerve roots, therefore complete removal of mass was not possible. Macroscopic de-bulking was done. The patient was relieved of pain after the surgery and there was no postoperative neurological deficit. Histopathology examination revealed islands of well-differentiated cartilage surrounded by diffuse proliferation of elongated to round cells arranged in fascicles & diffuse sheets having vesicular nuclei with small to large nucleoli, features suggestive of mesenchymal chondrosarcoma (Figure 2). Immunohistochemistry markers for tumor tissue were positive for S-100 and INL1, thus confirming the diagnosis. After consultation with the radio-oncologist, two weeks post-surgery the patient was sent for adjuvant radiotherapy. Patient was scheduled for follow up after six weeks but patient did not turn up, so we did a phone enquiry, where the daughter expressed the news of his death. The patient expired four

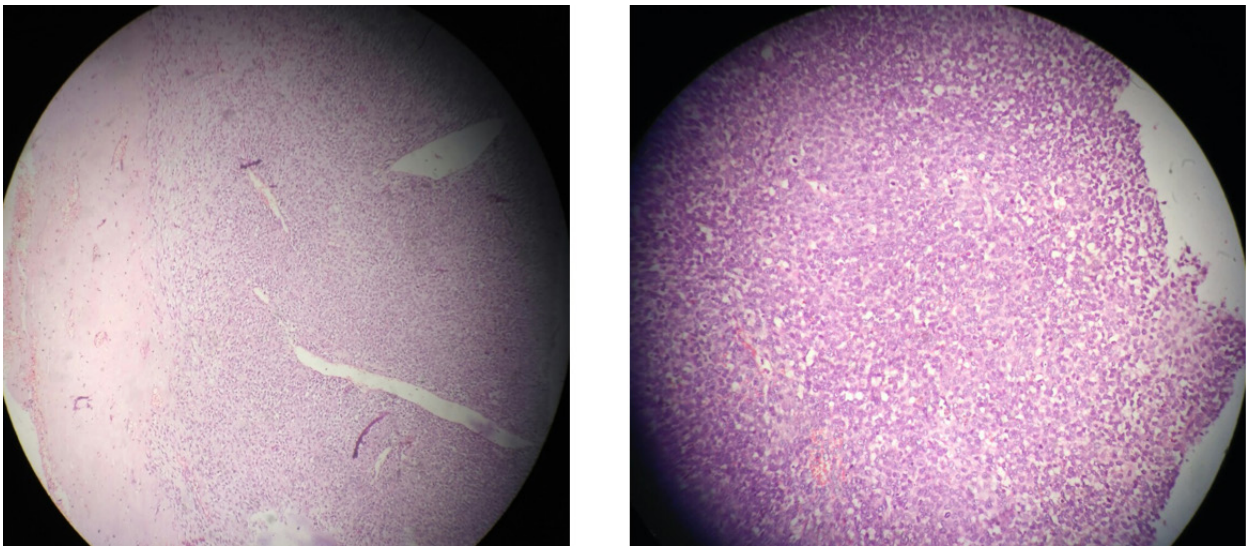

Fig 2. Histopathological photograph. Low power field (left) and High power field (right) which show islands of well differentiated cartilage surrounded by diffuse proliferation of elongated to round cells arranged in fascicles & diffuse sheets having vesicular nuclei with small to large nucleoli.

months postoperative and although the exact cause cannot be defined, most probable would be due to disseminated metastasis.

DISCUSSION

Chondrosarcoma is a primary malignant neoplasm of bone and soft tissue with both cartilaginous and undifferentiated round cell components and are histopathologically classified into three categories: classic, mesenchymal, and myxoid types.[1] MCS accounts for 3-10% of all primary chondrosarcoma. Although the majority of it originate from the bone, 33-50% can be detected in extra-skeletal sites. Extra-skeletal MCS most often involve the brain and meninges.[2] Most of the tumors arising from meninges are reported to be intracranial in location and very rarely within the spinal canal. Primary MCS can be found in any part of the spine, but they mostly occur in the thoracic segment. Extradural involvements of primary spinal meningeal MCS is again very rare, and majority have a dural attachment. A few publications described primary intraspinal dumbbell-shaped MCS.[3,4] In a case report by Harsh and Wilson[5] where they reviewed all the recorded cases till 1984, they found a total of 16 cases of MCS of the primary central nervous system where only five were intraspinal. Similarly, in another review of literature done by Obuchowicz et at[6] in 2010, they found 24 cases of intraspinal MCS from 1978 to 2010.

Though, younger age of presentation and female preponderance were observed in the past, no such differences are seen currently.[3,7] Our patient is a fifty-year-old male thus correlating with recent data. There are very few reports on sacral MCS and are mostly in paediatric age groups.[6] This might be the first report of sacral MCS at this age. Because of the intraspinal location of the tumor, the patient presented quite early with features of radiculopathy and pain. Intraspinal lesions are detected earlier than other soft tissue MCS because of earlier symptoms of radiculopathy and pain akin to the site.[3,4,7] MRI with or without gadolinium enhancement is the imaging modality of choice for intraspinal tumors, but there are no pathognomonic signs in the intradural chondrosarcomas. On T1-weighted images, the signals are usually isointense to the normal spinal cord while T2-weighted images show a high intensity or isointense. The overall signal intensity is homogeneous with both short and long repetition times, but if there is calcification, which is pathognomonic in chondrosarcoma, it appears as various signals in MRI, depending on the degree of differentiation. Images obtained after administration of gadolinium shows strong homogenous enhancement.[4] Our case also had a similar picture with enhancement after gadolinium but no calcifications were present. However, due to the nonspecific nature of imaging examination to identify MCS, the hemangioblastoma, neurofibroma, and schwannoma should be kept as differentials.[4]

The MCS is a hard or fish-meat like, grayish-white or grayish-red mass, mixed with calcifications sometimes.[3] During operation, we also encountered a fluffy fish meat like grayish-white mass. Histologically, most MCSs exhibit a biphasic pattern of islands of cartilage and areas of neoplastic small blue round cell component. The exact histogenesis of intraspinal chondrosarcomas is obscure because these lesions are generally associated with cartilage.[8] It is postulated that chondrosarcomas arise from the dura because it has the periosteal component over the spinal extradural vault. These chondrosarcomas may arise from embryonic rest cells of cartilage within the dura. The tumor after arising from the dura invades the surrounding extradural space and neural foramina. Diagnostic immunohistochemistry findings include positivity of the mesenchymal portion for vimentin, Leu[7], and CD99 and positivity of the cartilaginous regions for S100 protein which helps to differentiate it from Ewing's sarcoma and hemangiopericytoma.[4]

The prognosis is poor because they tend to recur locally, metastasize, and are resistant to chemotherapy or radiation therapy. Radical surgery with complete removal of the tumor is considered the best choice of treatment.[9] The tumor was present extensively in extradural spaces and delineation with normal tissues was not possible so we could not resect the tumor radically. Even though some literatures conclude the effect of irradiation on the prognosis, most authors agree that radiotherapy is recommended in cases of partial resection.[7,10] We treated the patient with postoperative radiotherapy since the tumor was removed partially, but the patient expired in few months.

CONCLUSION

Primary spinal mesenchymal chondrosarcoma should always be kept in differentials of any patient presenting with radiculating back pain and a mass lesion in MRI. As the tumor has an aggressive course early diagnosis is the key to successful treatment. Low threshold for MRI evaluation for suspected patient can help in earlier diagnosis. Spinal mesenchymal chondrosarcomas have a poor response to chemotherapy or radiotherapy and so, complete resection of the tumor should be done.

CONFLICT OF INTEREST

None declared.

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