Mesenchymal chondrosarcoma of the cervical spine: A case report

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CASE REPORT

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Abstract

Mesenchymal chondrosarcoma (MCS) is a rare tumour accounting for less than 1% of all chondrosarcomas. We report here, the case of a 30-year-old female who presented with neck pain, weakness and tingling and numbness starting on the right side and later involving all four limbs. MRI revealed an ill-defined neoplastic lesion showing specks of calcification and arising from the right half of C2 and C3 cervical vertebrae. Microscopy showed an admixture of well differentiated cartilage showing focal calcification and spindly cell areas having a hemangiopericytomatous pattern.

Key Words
Chondrosarcoma, mesenchymal, spine

Background

MCS is an uncommon malignant chondrogenic neoplasm with an overall poor prognosis. It represents about 1% of all chondrosarcomas and affects all ages with a peak occurrence in the second decade of life and has no sex preponderance. A peculiarity of this neoplasm is that approximately one-third of cases develop outside the bone including a significant number arising in the meninges.[1] In spite of being rare, this tumour should be considered in the differential diagnosis of spinal tumours[2] along with other tumours common to this location and age group such as Ewing's sarcoma and osteosarcoma.[1] Metastases to regional and distant lymph nodes in addition to other bones are seen in MCS. This peculiarly strong inclination of unusual metastatic dissemination further strengthens and justifies clinical separation of MCS from other chondrosarcomas.

Case details

A 30-year-old female presented with a two-month history of neck pain, progressive weakness, tingling and numbness starting on the right side and later involving all four limbs. MRI (T_1 weighted mid-sagittal section) revealed a large irregularly marginated mass lesion on the right half of C2, C3 vertebral bodies (Figure 1). The mass showed specks of calcification. Anteriorly, it extended into the right prevertebral and right paravertebral region from C1 to C4 causing extrinsic compression and deviation of the pharynx. Posteriorly, the intraspinal epidural component present on the right side caused spinal cord compression and deviation to the left side. Multiple enhancing enlarged lymph nodes were noted in the right cervical posterior triangle. A diagnosis of chondrosarcoma was considered. The mass was surgically excised. We received fragmented soft translucent greyish white material admixed with bony spicules. Microscopy revealed a tumour composed of islands of well differentiated cartilage surrounded by ovoid to spindly cell areas exhibiting hemangiopericytomatous pattern (Figure 2). Focal calcification was seen in chondroid areas. Individual cells were ovoid to spindly with bland nuclear features. Histology favoured the diagnosis of mesenchymal chondrosarcoma.

Confidentiality

All identifying information has been removed from the material presented in this report.

Discussion

MCS is a malignancy that is classed as a chondrosarcoma due to the focal appearance of cartilage alternating with undifferentiated stroma.[1] It was first recognised by Lichtenstein and Bernstein in 1959.[3] It is a rare undifferentiated neoplasm of bone and soft tissue.[4] It is slightly more common in females than in males and occurs at an average age of 25 years. However some cases have

been reported in elderly patients as well.[5] Extra-skeletal MCS affects young patients while those involving bone affects older patients. MCS presents as a mass or swelling with pain of varying degrees of severity at the site involved and the lesional size varying from 4–18cms.[5]

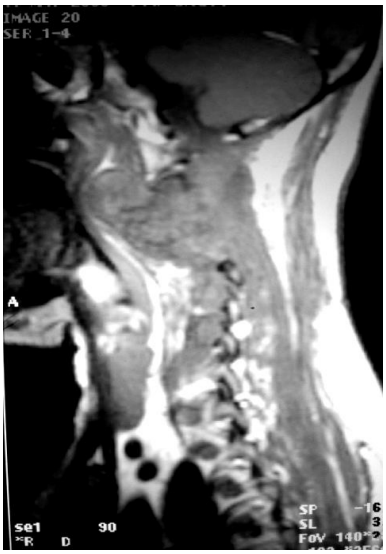

Figure 1: MR image showing an irregularly marginated mass lesion arising from the right half of C2, C3 vertebra

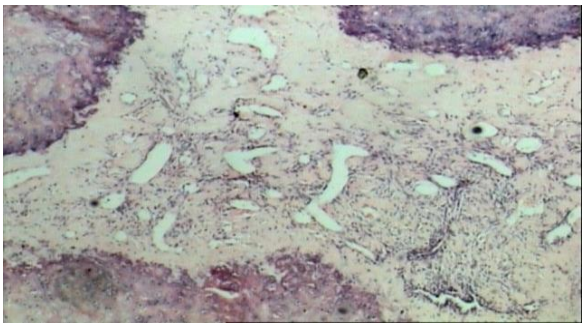

Figure 2: Photomicrograph showing well-differentiated cartilaginous component and highly cellular spindle cell area showing hemangiopericytomatous pattern (H&E 40X)

Primary chondrosarcoma of the axial skeleton is uncommon and usually affects the posterior part of the vertebral body.[6] Spinal chondrosarcoma is characterised by the radiological evidence of destruction of the surrounding bone structure and mottled calcification.[7] Microscopically they have a bimorphic histologic appearance composed of highly cellular undifferentiated areas and areas showing well differentiated cartilage. These two elements are arranged either as distinct areas or they can be relatively mixed up. The cellular areas have the appearance of a small round blue cell tumour and may have hemangiopericytomatous areas. Despite the undifferentiated nature of this component, pleomorphism and mitotic activity are inconspicuous. [8] It has been proposed that MCS represents a neoplastic caricature of embryonal endochondral osteogenesis.[8] Jacobson postulated that MCS is one morphologic type of bone tumour that he proposes to call polyhistioma. He defines it as a malignant neoplasm whose basic cells are small and round like those of Ewing's sarcoma but that differentiates into various mesenchymal structures such as bone and cartilage and sometimes even into epithelial structures. Immunohistochemically small cell component is positive for vimentin, CD99, Leu7 but not for S100 protein which is found instead in chondroid areas. There is also nuclear immunoreactivity for Sox 9, a master regulator of chondrogenesis.[8] MCS is a fast growing tumour and is known to spread to the lymph nodes, lungs, soft tissues and other major organs. The prognosis for patients with MCS is usually poor and long-term follow-up is essential because local recurrence or metastases are sometimes encountered even after more than 20 years.[9] Patients without evidence of metastasis generally have a better prognosis.[5] The modalities for treatment include surgical resection followed by chemotherapy (adjuvant chemotherapy)[10] and neo-adjuvant chemotherapy (chemotherapy followed by surgical resection).[11] Radiation may be used after complete resection to treat residual tumour cells[12] and also when complete resection is not possible. The period for relapse as well as disease progression is protracted.[5] Our patient underwent surgical resection and was referred to an advanced centre for further management by chemotherapy and radiotherapy. The patient was, however, lost at follow-up.

Currently, the histological diagnosis of bone tumours is nearly exclusively based on conventional histology. In the future, the use of markers of mesenchymal cell differentiation may also have an impact on the differential diagnosis in critical cases.

This case was presented to highlight the features of MCS as a spinal tumour, the diagnosis of which may be missed because of its rarity and lead to increased patient morbidity due to its protracted clinical course.

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CONFLICTS OF INTEREST
The authors declare that they have no competing interests.