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Rare Cases of Cranial and Spinal Mesenchymal Chondrosarcoma

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Abstract

Mesenchymal chondrosarcoma is a rare, aggressive subtype of chondrosarcoma with a widespread anatomical distribution in bone, soft tissue and intracranial locations. It accounts for 2 to 10 % of all chondrosarcomas. Intraosseous mesenchymal chondrosarcomas are seen in craniofacial bones, ribs, ilium, vertebrae and lower extremities. Intracranial and spinal mesenchymal chondrosarcomas are rare. Histologically it has a biphasic morphology consisting of atypical cartilage and small round blue cells. We report 2 cases of mesenchymal chondrosarcoma in unusual locations: the petrous part of the temporal bone and cervical spine.

Keywords: Mesenchymal Chondrosarcoma, Cranial tumour, Spinal tumour, Hemangiopericytoma like pattern

Introduction

Mesenchymal chondrosarcoma is a rare malignant mesenchymal tumour with well differentiated cartilage component. Lichtenstein and Bernstein first described it in bone in 1959 (1). Mesenchymal chondrosarcoma accounts for only 2% to 10% of all chondrosarcomas (2). Intraosseous mesenchymal chondrosarcomas are seen in craniofacial bones, ribs, ilium, vertebrae and lower extremities (3). Mesenchymal chondrosarcoma of the spine as a primary lesion is extremely rare. These are highly aggressive tumours with an overall 5-year survival rate of 60%.

Aim

We present 2 cases of mesenchymal chondrosarcoma located at the cervical spine and petrous part of the temporal bone.

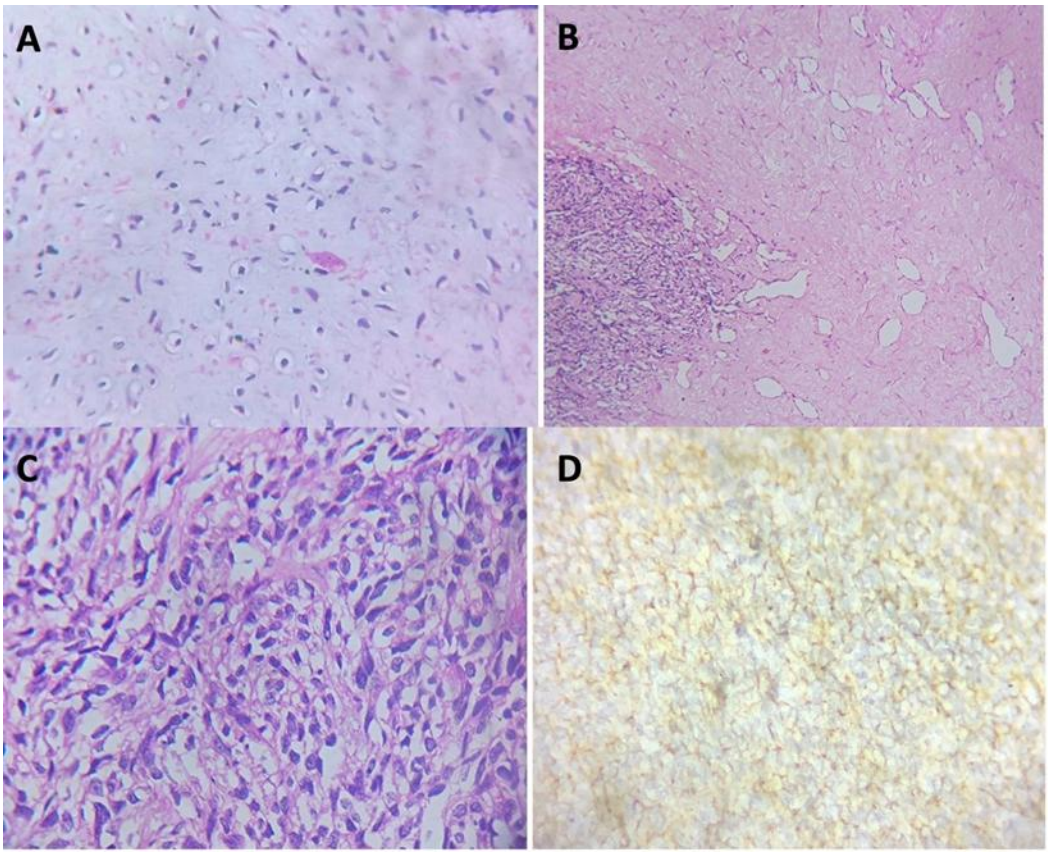
Cases

Case No:1

A 28-year-old male presented with complaints of bilateral upper limb and lower limb weakness for the past 6 months. An MRI was taken and it showed a well-defined contrast-enhancing lesion involving the left anterior and posterior region of C7 vertebrae with neural foraminal widening. The radiological impression was mesenchymal tumour. A guided biopsy from the tumour showed a spindle cell neoplasm with hemangiopericytoma like pattern. The tumour was surgically removed.

We received a grey-white nodular mass measuring 4.5*2.5*2 cm. The cut section showed myxoid and haemorrhagic areas. Microscopy showed a neoplasm with a biphasic pattern with islands of hyaline cartilage separated by sheets of small blue cells in hemangiopericytoma like pattern. Hyaline cartilage showed mild nuclear atypia. Immunohistochemical study showed membranous positivity for CD99 and cytoplasmic positivity for Vimentin. Hence a final diagnosis of Mesenchymal Chondrosarcoma was made.

Figure 1 (A) Islands of cartilage with mild atypia, (B) Staghorn vessels (C) Small to medium-sized poorly differentiated cells, (D) CD99 membranous positivity



Case No: 2

A 26-year-old female presented with complaints of neck pain radiating to her right hand, right sided hearing loss and tinnitus for the past 6 months. On clinical examination, Right LMN facial palsy and right sided moderate sensorineural hearing loss were noted. MRI Brain revealed a large extra-axial lesion in the right temporal bone which was hypo-intense on T1 and hyper-intense on T2 and FLAIR. The tumour was encasing the internal carotid artery. The features were suggestive of chondrosarcoma.

The patient underwent craniotomy. Intraoperatively a firm, moderately vascular tumour involving the petrous part of the right temporal bone was noted. A frozen section from the tumour showed fragments of cartilage and poorly differentiated cells with scant cytoplasm and hyperchromatic nucleus.

We received multiple grey-white tissue bits aggregate measuring 5*4.5*2. The Cut section showed grey-white and haemorrhagic areas. Microscopy showed a neoplasm composed of small to medium sized poorly differentiated cells with scanty cytoplasm, high nucleo-cytoplasmic ratio arranged in sheets, admixed with well differentiated hyaline cartilage. Foci of high-grade cytological atypia were noted in the islands of cartilage. Areas of necrosis, calcification and atypical mitosis were also noted. Immunohistochemistry showed S100 positivity in cartilaginous areas and strong membranous positivity for CD99. The final diagnosis was Mesenchymal Chondrosarcoma, petrous part of right temporal bone.

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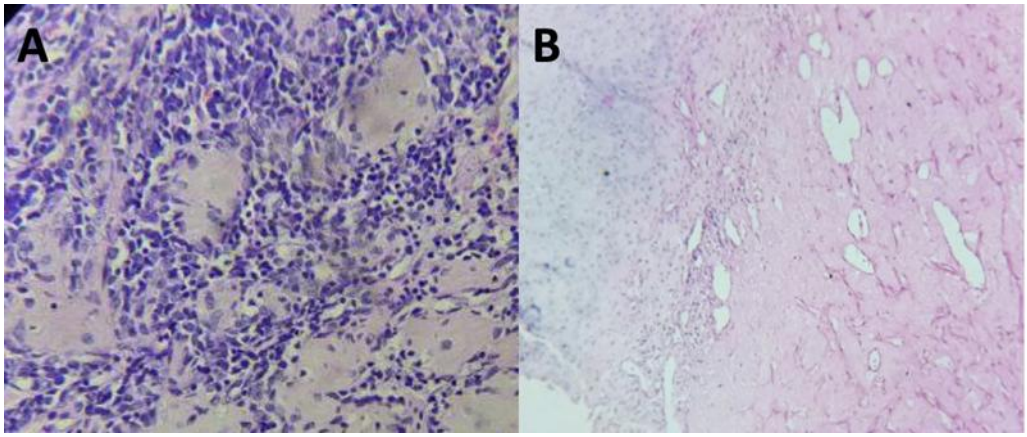
Figure 2: (A) Small round blue cells, (B) Islands of cartilage and staghorn vessels.



Discussion

Mesenchymal chondrosarcoma is a rare subtype of chondrosarcoma with widespread anatomical distribution in bone, soft tissue and intracranial locations. Within the bone, the neoplasm is located frequently in the medulla. Although most of these tumours are thought to originate in the bone, 50% of reported cases arise from extraosseous locations, including the brain and meninges (4). Mesenchymal chondrosarcoma of the spine as a primary lesion is extremely rare. It is most commonly seen in young adults and children.

Mesenchymal chondrosarcoma is different from typical chondrosarcomas in various aspects. Mesenchymal chondrosarcomas are slightly more common in females and occur in the nervous system in patients with age 20 to 30 and in soft tissues in patients aged 40 or older, whereas classical chondrosarcomas are commonly seen in middle-aged to elderly males. Mesenchymal chondrosarcoma tends to be more aggressive.

The classical clinical symptoms of Mesenchymal chondrosarcoma include pain and swelling (5). Due to the nonspecific symptoms and findings, the diagnosis is often delayed. Radiographically, mesenchymal chondrosarcoma looks like an osteolytic neoplasm showing radiolucent lesions with matrix calcification in arches, rings, and stippled patterns (6). No unique radiographic features specifically differentiate mesenchymal chondrosarcoma from other chondrosarcomas (7). HEY1 and NCOA2 gene fusion is found in almost all mesenchymal chondrosarcomas and is highly specific.

Grossly the tumours are grey-white to pink, firm to soft, circumscribed masses. Microscopy shows the characteristic biphasic pattern composed of a solid mass of undifferentiated small round or spindle cells and islands of chondroid tissue (8). Staghorn or pericytoid vascular pattern is also characteristic. On immunohistochemistry, tumour cells can be positive for S100, CD99 and SOX9. EMA, Myogenin and desmin may show aberrant expression. This highly aggressive tumour tends to metastasize to lymph nodes, lungs and other bones. Ewing's sarcoma and hemangiopericytoma are important differential diagnoses.

Surgery, adjuvant chemotherapy and radiotherapy are the treatment options. Radiotherapy aids in improving people's survival when complete resection is impossible (9).

Conclusion

Cranial and spinal mesenchymal chondrosarcomas are rare neoplasms that are challenging in terms of diagnosis and surgical management. The diagnostic diagram includes radiology, histopathology with immunohistochemistry and molecular analysis. The present treatment strategy includes maximal resection followed by radiotherapy.

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