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Case Report

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The curious case of a paraspinal mass

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ABSTRACT

Chordomas are rare, slow-growing malignant neoplasms arising from embryonic notochord remnants, most commonly found in the sacrococcygeal region and skull base. Cervical chordomas are exceptionally rare, accounting for only 6% of cases. We present a case of a 64-year-old man with cervical chordoma at the C5 vertebral level, manifesting as cervical myelopathy with limb numbness and bowel/bladder dysfunction. MRI and CT angiogram revealed a well-lobulated, lytic mass involving C5–C6, with encasement of bilateral vertebral arteries. Initially suspected as a metastatic lesion, surgery was undertaken without biopsy due to high risk of neurovascular injury. A C4–C6 corpectomy and tumor resection were performed via an anterior transcervical approach. Histopathology and immunohistochemistry confirmed the diagnosis of chordoma. Postoperative recovery was uneventful, with improved neurological status and no recurrence on follow-up MRI at 12 months. Chordomas can mimic other spinal lesions radiologically, complicating diagnosis. MRI, especially with gadolinium contrast, remains the imaging modality of choice. Treatment involves radical surgical resection, which is the most significant prognostic factor, often supplemented by adjuvant radiotherapy. Due to their resistance to conventional chemotherapy and high recurrence rates, emerging therapies such as tyrosine kinase inhibitors and molecular targeted agents (e.g., imatinib, sorafenib) are under investigation. Despite advancements, management remains challenging due to proximity to critical structures and limited systemic therapy options. Multidisciplinary planning is essential for optimal outcomes. This case highlights the diagnostic and therapeutic complexities of cervical chordoma and underscores the importance of individualized, team-based treatment approaches.

Keywords: Chordoma, Cervical, Primary bone tumors

INTRODUCTION

Chordomas are uncommon malignant tumors, with an incidence rate of less than 0.1 per 100,000 individuals, and they are most frequently seen in middle-aged men. They represent approximately 1–4% of all primary bone tumors, with a predilection for the sacrococcygeal area and the skull base.

Only about 6% of chordomas are located in the cervical spine, typically presenting in the midline. These tumors are characterized by slow growth and often remain asymptomatic until they enlarge enough to involve adjacent vascular and neural structures. Advanced cases generally have a poor prognosis due to the tumor's tendency to locally recur and invade surrounding bone and soft tissue.

Traditional chemotherapy has shown limited effectiveness, prompting interest in molecularly targeted treatments, particularly tyrosine kinase inhibitors. Nevertheless, the cornerstone of treatment remains complete surgical removal of the tumor followed by postoperative radiotherapy.[1]

CASE REPORT

A 64-year-old man diabetic, hypertensive and hyperthyroid was admitted to our hospital with complaints of generalised weakness and numbness in both upper limbs and bilateral lower limbs for a year with bowel /bladder impairment. (Modified Nuricks Grade II, MJOA 12). On clinical examination, he had features of cervical myelopathy with Grade 5 power in all limbs with hyperreflexia and bilateral handgrip weak with bilateral plantar extensor. MRI C-Spine was suggestive of a well-lobulated lesion centered on C5 vertebral body extending to C4 and C6 with spinal canal narrowing and cord impingement.

CT-C-spine with angiogram showed expansile lytic destruction of the C5 Vertebral body with encasement of bilateral vertebral arteries at C5 and C6 with a well-defined soft tissue density of 6.7×6.2 cm surrounding C5-C6 vertebral bodies with erosion of C5-C6 vertebral bodies and bilateral vertebral foramen and transverse process. Metastatic workup was done for the primary lesion with a preoperative diagnosis of metastasis but was inconclusive. An ultrasound-guided FNAC was done which showed only inflammatory cells. Based on its localization, and neuroradiological and clinical features, the tumor was preliminarily diagnosed as a metastatic lesion.

The patient was qualified for surgery without prior preoperative biopsy, since performing biopsy carried a risk of neurological deficit, with damage to adjacent vascular structures. Surgery was performed through an anterior transcervical approach procedure with a longitudinal incision from C2 to C6, along the medial border of sternocleidomastoid. The cervical neurovascular bundle and vertebral artery were identified and preserved. Then, local structures were mobilized and pulled to the sides, exposing the tumor. Intraoperatively, a firm and well-encapsulated mass was seen along C4 to C6 under the microscope, which was carefully evacuated in fragments. C4, C5, C6 corpectomy with cervical cage fusion and biopsy was done.

The patient's postoperative course was uneventful with the patient improving clinically and ambulant with Grade 5 power in all limbs (Modified Nuricks Grade II, MJOA 15), with discharge from the hospital after four days. Incidentally, the diagnosis of chordoma was established by histopathological features.

The histopathological image revealed round neoplastic cells with a centrally located nucleus and pale eosinophilic, vacuolated cytoplasm, arranged in cords separated by fibrovascular bands and myxoid matrix. Nuclear pleomorphism was heterogeneous, but mitosis was rare (Ki67 <1%). An immunohistochemical study revealed positive reactivity for EMA, AE1/AE3, S100, and negative for GFAP, which indicated notochordal origin and confirmed the final diagnosis of chordoma

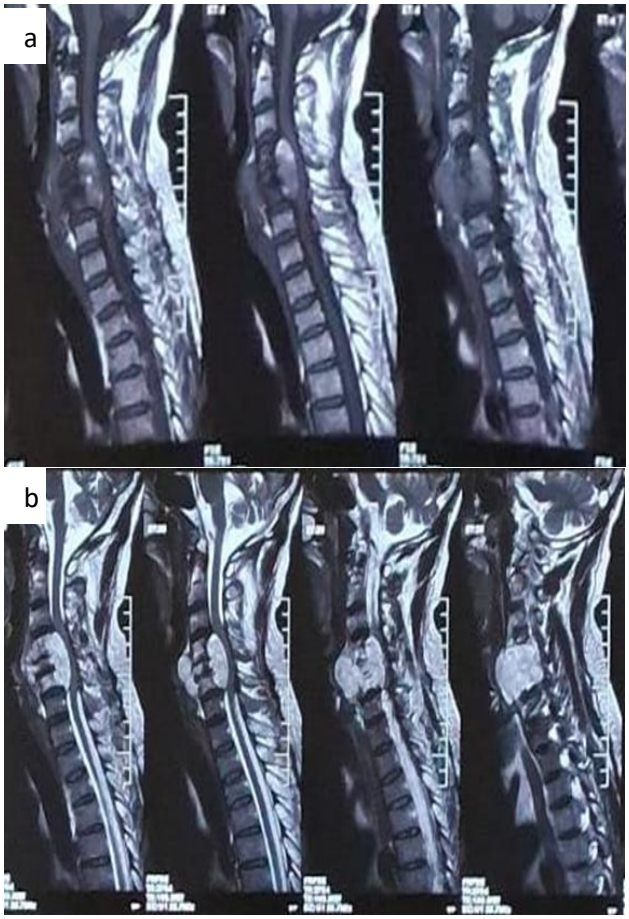

Figure 1: MRI cervical spine T1 and T2 weighted image sagittal view.

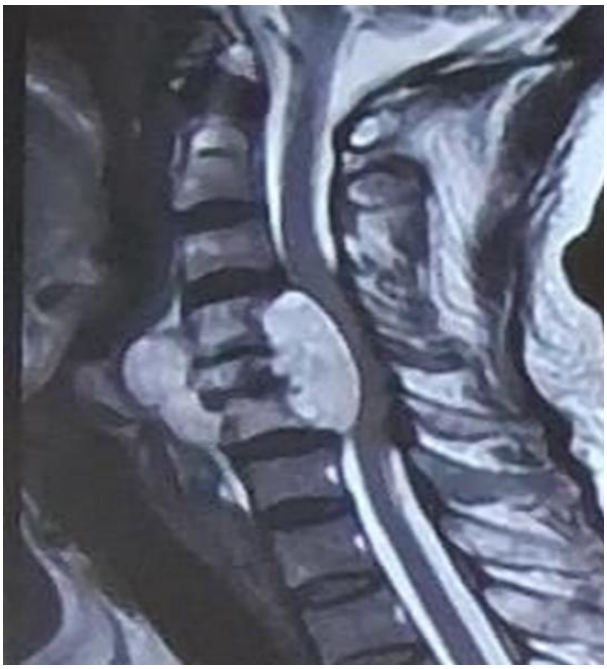

Figure 2: MRI C-Spine-a well-lobulated lesion centered on C5 vertebral body extending to C4 and C6 with spinal canal narrowing and cord compression.

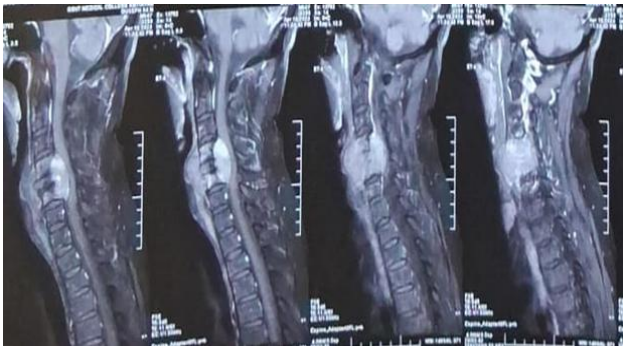

Figure 3: MRI cervical spine with contrast.

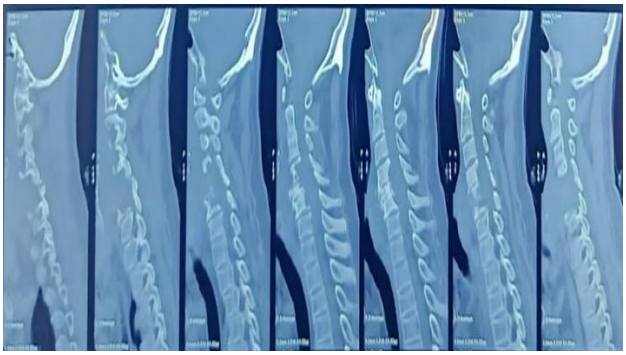

Figure 4: CT cervical spine-sagittal view.

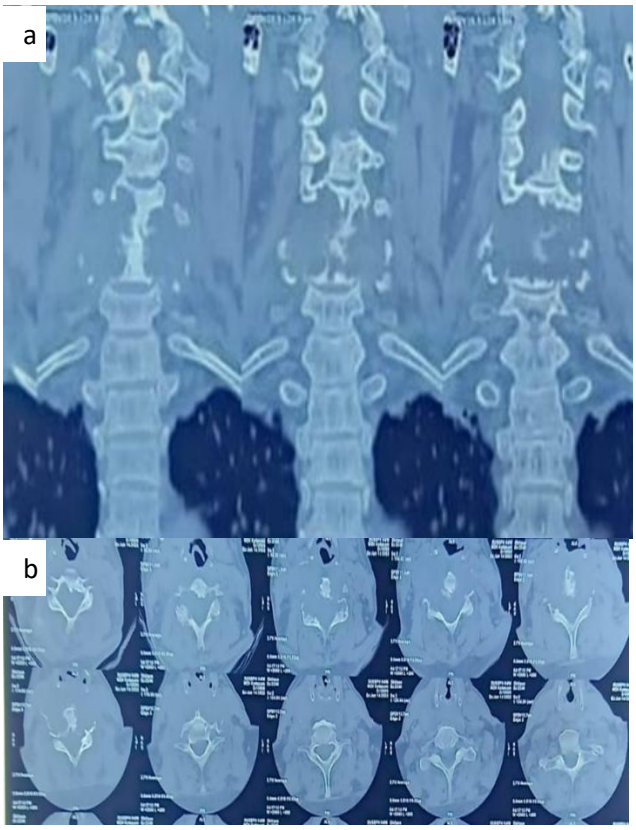

Figure 5: CT cervical spine coronal view and CT cervical spine axial view. CT showing C4, C5, C6 vertebral body destruction in axial coronal and sagittal views.

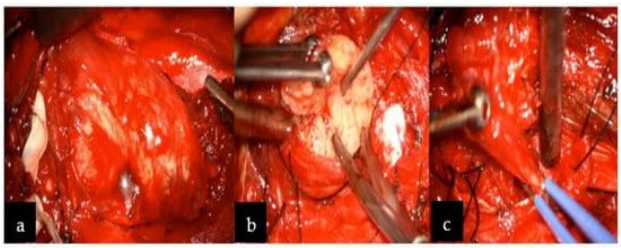

Figure 6: Intraoperative image revealing: (a) the paravertebral well-capsulated tumor on the right side at the C2–C4 level, (b) after debulking, and (c) complete excision was achieved. Below the tumor, vertebral artery was identified (not indicated in the image) and above the tumor, the hypoglossal nerve was visualized.

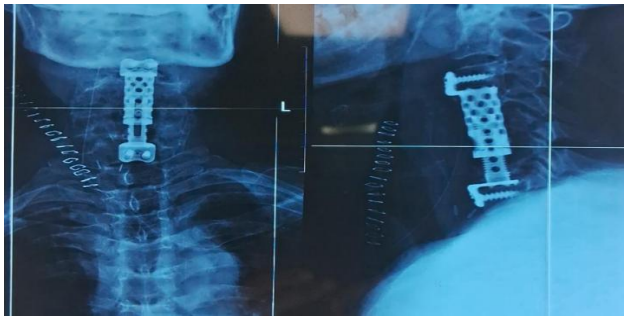

Figure 7: Post op X-ray-C4, C5, C6 corpectomy with cervical cage fusion.

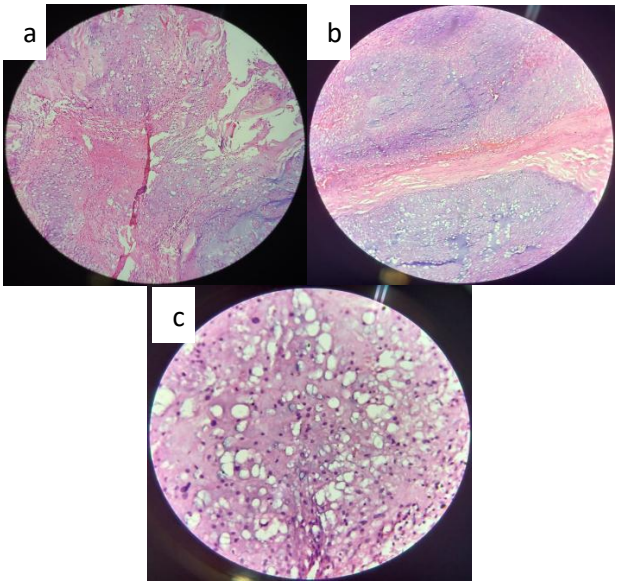

Figure 8: Extracellular myxoid matrix 2-cords of epitheloid cells 3-physaliphorous cells with bubbly cytoplasm. a-Extracellular myxoid matrix b-cords of epitheloid cells c-physaliphorous cells with bubbly cytoplasm. Hematoxylin–eosin stain, neoplastic cells with pale eosinophilic, cytoplasm vacuolated, arranged in cords separated by fibrovascular bands and myxoid matrix. Immunoreactivity for AE1/AE3, S100, and negative one for GFAP.

DISCUSSION

Chordomas are a unique category of primary bone malignancies that originate from residual embryonic notochord tissue, typically found along the central axis, extending from the skull base to the sacrum. This case is notable due to the uncommon cervical location and the diagnostic challenges posed during preoperative assessment via imaging and clinical evaluation.[2]

Radiological characteristics

Chordomas are typically classified into four variants: classical and chondroid types-which are low-grade but locally invasive-and the more aggressive poorly differentiated and dedifferentiated forms. Imaging often reveals bone destruction and lytic changes, sometimes with sclerosis extending into the spinal canal. Initial evaluation usually begins with radiographs, which may show bony erosion with abnormal calcification. CT imaging better characterizes hypodense, calcified lesions and extent of bony involvement, with sclerosis observed in up to 60% of cases. MRI remains superior in assessing tumor extent and its relationship with surrounding structures. On MRI, chordomas typically appear hypointense on T1-weighted images with post-contrast enhancement, and hyperintense on T2-weighted sequences due to internal degeneration or calcification.[3]

Spinal chordomas may present as large paraspinal masses that compress or invade neural structures and enlarge neural foramina. Their imaging features can resemble other pathologies such as schwannomas, neurofibromas, metastases, chondrosarcomas, meningiomas, or hematologic malignancies. Differentiating chordomas from these entities is often complex. Notably, chondrosarcomas tend to involve the posterior spinal elements, whereas chordomas more commonly affect the vertebral body. Intralesional calcification may suggest chordoma, but multifocal spine involvement typically excludes it. Histology is critical, and markers like brachyury are used to distinguish chordomas from mimickers.[4]

Classification systems

Chordomas are staged using systems like the Enneking classification, which evaluates tumor grade and extent, and the Weinstein–Boriani–Biagini (WBB) classification, which provides anatomical context for spinal tumors and guides surgical planning.[4]

Treatment strategies

Management strategies have included combinations of surgery and radiotherapy. Complete surgical excision with clear margins offers the best chance of long-term disease control, although it is often difficult due to the tumor's proximity to vital structures. The standard approach includes aggressive surgical resection followed by adjuvant radiation therapy, improving 3- to 5-year survival to around 70%. If full resection is not feasible, partial debulking followed by radiotherapy may be pursued.[5]

Surgical margins are a critical prognostic factor. En bloc resection with preservation of the tumor capsule and negative margins reduces recurrence risk. Intraoperative spillage should be avoided, and margin status must be assessed pathologically. Surgical planning must consider tumor location, adjacent neurovascular structures, and the patient's neurological function to maximize resection while minimizing morbidity. Although preoperative biopsy is recommended when chordoma is suspected, it was avoided in this case due to high risk of neurological or vascular injury.[6]

Role of radiotherapy

Cervical chordomas pose challenges for radiation therapy due to their location near the spinal cord and vertebral arteries. While photon or proton beam therapies offer precision, their effectiveness is enhanced when delivered early in treatment, ideally following primary resection. Some evidence supports preoperative radiotherapy as potentially more beneficial than postoperative treatment. Proton therapy is emerging as the preferred modality for its tissue-sparing advantages.

Given the high rate of local recurrence, especially after subtotal resections, conventional radiotherapy alone is not curative. This limitation has prompted exploration into targeted molecular therapies as adjunctive or alternative treatments.[7]

Emerging therapies

Research is ongoing into therapies targeting specific pathways implicated in chordoma growth, such as EGFR and PDGFR. Agents like imatinib, erlotinib, sorafenib, lapatinib, and sirolimus have shown promise in early studies. Additional options under investigation include CDK4/6 inhibitors like palbociclib, and immune checkpoint inhibitors such as avelumab and nivolumab.[8,9]

CONCLUSION

Due to their rarity and complex presentation, chordomas require individualized treatment planning. Systemic therapy options remain limited, and most current strategies focus on surgical resection with or without radiotherapy. Targeted therapy represents a promising frontier, particularly for recurrent disease. However, further research and clinical trials are necessary. Optimal management depends on detailed preoperative planning by a multidisciplinary team to ensure maximal resection and minimize recurrence, offering the best chances for favorable outcomes.

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