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Chordoma of low cervical vertebrae –A rare site- Case report

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Abstract

Chordoma is an uncommon and malignant bone tumor that originates from remnants of the embryonic notochord. To date, only nine cases of lower cervical chordomas have been documented globally. This study aims to highlight an extraordinary case involving a 14-year-old female with chordoma affecting the fifth to seventh cervical vertebrae.

Keywords

Chordoma, Cervical vertebrae, bone neoplasm

Introduction

Chordoma is a rare and malignant bone tumor originating from remnants of the embryonic notochord (1-3). Approximately 85% of chordomas develop in the sacrum, followed by the spheno-occipital area (35-45%), with the least common occurrence in vertebral bodies (15-20%) (4, 5). Annually, chordomas arise in only 0.08 per 10,000 cases, predominantly affecting males with a peak incidence in the fifth decade of life (6). Despite being classified as a benign neoplasm, chordomas possess significant malignant potential due to their high recurrence rate. Cases of chordomas arising in the cervical spine are exceptionally rare, particularly in the lower cervical region (C5, C6, and C7) (2). To date, only nine such cases have been reported in the literature. The present study is an endeavor to report an extraordinary case of a 14-year-old female with a chordoma in the fifth to seventh cervical vertebrae.

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Case report

A 14-year-old female was admitted to the Neurosurgery Department at the National Institute of Medical Science and Research with sudden urinary retention for two days, followed by the rapid onset of weakness in both upper and lower limbs. Neurological examination revealed slight weakness in the flexion of both upper limbs (4/5 power) and moderate weakness in their extension (3/5 power). The left lower limb exhibited significant weakness in flexion (2/5 power), while the right lower limb showed extreme weakness in extension (1/5 power). There was no significant family medical history. Despite steroid treatment, her symptoms did not improve.

A plain X-ray revealed anterior tracheal displacement without vertebral collapse. T1-weighted MRI in the sagittal plane identified an anterior soft tissue mass at the C5 level extending to the C7 level, displaying low signal intensity and causing anterior displacement of the trachea (Figure 1). Routine laboratory tests, including a tuberculin test, were normal.

The patient underwent surgery to remove the mass, which was subsequently sent to the Department of Pathology for histopathological examination in a 10% formalin solution. The specimen comprised multiple soft tissue pieces, pale brown in color, measuring 2x1 cm, with a gelatinous cut surface. Histopathological examination revealed cells forming short chords and lying singly. The cells were epithelioid with abundant clear to eosinophilic cytoplasm and a vacuolated appearance (physaliphorous cells). There was mild nuclear pleomorphism and brisk mitosis within an extracellular myxoid matrix (Figure 2).

Immunohistochemistry (IHC) performed on the tissue showed that the tumor cells were EMA and S100 positive (Figures 3 and 4), CK7 negative, and focally positive for CK19 (Figures 5 and 6). These findings confirmed the diagnosis of chordoma.

The surgery was uneventful, and the patient was discharged a few days later with a recommendation for follow-up to receive further radiotherapy.

Discussion

Chordoma is a rare, slow-growing, but locally invasive neoplasm that accounts for less than 5% of all bone tumors (6). Originating from the notochord, the embryonic precursor to the vertebral

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column, chordomas typically develop in the midline from the clivus to the sacrum (1). Although they predominantly occur in the sacral area, chordomas can occasionally present in atypical locations such as the transverse process of a vertebra (7).

Patients usually present in their fifth to sixth decades, with a mean age of 55 years. However, rare cases in younger individuals have been documented, including a 10-month-old patient with a clival chordoma reported by Backer et al., (8) and a one-month-old infant with a thoracic vertebral chordoma described by Coffin et al (9). There is a male predominance of approximately 2:1 (1-4, 6-10).

Grossly, chordomas are lobulated, gray, and soft, well-demarcated but with elusive bone margins. While generally slow-growing, they are locally aggressive and can metastasize to the lungs, lymph nodes, bone, skin, and brain (9-13). Radiologically, vertebral destruction or lytic lesions with occasional sclerotic changes are common, and calcifications are seen in 15-18% of cases (1, 13-14). MRI is preferred over CT for its superior delineation of soft tissue tumor extension, invasive adherence, and site of origin. CT scans typically show a low attenuated hyperdense soft tissue mass with calcification, lytic lesions, and local destruction (15).

Differential diagnoses include chondrosarcoma, metastatic tumors, and chondromas (12,14, 16). Cervical chordomas are rare. A study by Utne et al. in 1955 reported a 4.1% incidence of cervical chordomas (17), while Dahlin et al. found a 7% incidence (18). Sundersan et al. documented five cervical tumors out of 54 cases. Cervical chordomas most commonly occur in the second vertebra, but our case involves the fifth, sixth, and seventh cervical vertebrae, which is exceptionally rare.

The most common presenting complaint is local pain without neurological signs unless nerve root involvement causes compression, complicating diagnosis. Myelopathy signs appear late, and anterior tumor extension often causes dysphagia and breathing difficulties. Sundersan et al. reported that two out of five cases presented with retropharyngeal masses (1).

Since chordomas respond poorly to radiation and chemotherapy, surgical resection remains the primary treatment. While resection in the sacrococcygeal area is typically curative, cervical chordoma resections are rarely successful due to anatomical constraints (19).

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Conclusion

In conclusion, chordomas are rare, locally invasive neoplasms originating from embryonic notochord remnants. While most commonly located in the sacral region, these tumors can occasionally occur in the cervical spine, as demonstrated by the exceptionally rare case involving the fifth, sixth, and seventh cervical vertebrae in a 14-year-old female. The diagnostic challenge posed by their atypical presentation and the complexity of their anatomical location necessitates a high index of suspicion and the use of advanced imaging techniques, particularly MRI, for accurate diagnosis. Given the poor response of chordomas to radiation and chemotherapy, surgical resection remains the cornerstone of treatment, although it is often complicated by the anatomical constraints of the cervical spine. Continued research and documentation of such rare cases are crucial for enhancing the understanding and management of this uncommon and challenging tumor.

Conflict of interest

None as stated by authors

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None as stated by author

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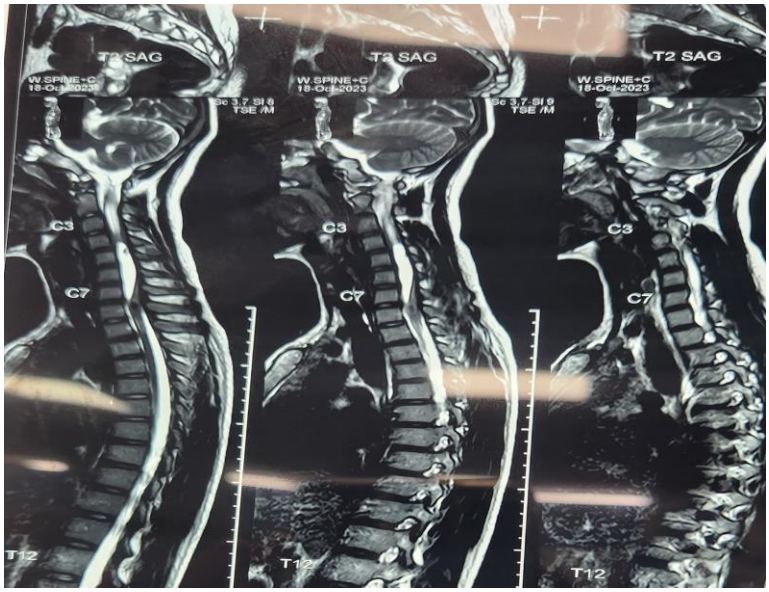

Figure 1- MRI showing anterior soft tissue mass which was cystic at C5 level extending till C7 level with low signal intensity which displaced trachea anteriorly

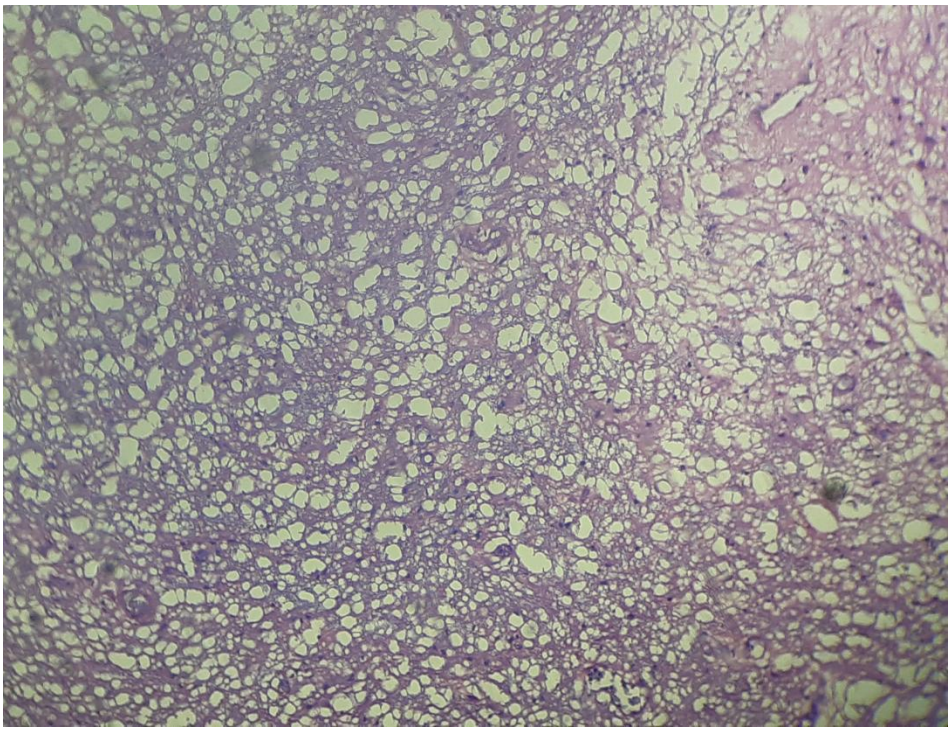

Figure 2 (H&E 40X) Cells forming short chords and lying singly. Cells were epithelioid with abundant clear to eosinophilic cytoplasm having a vaculated appearance

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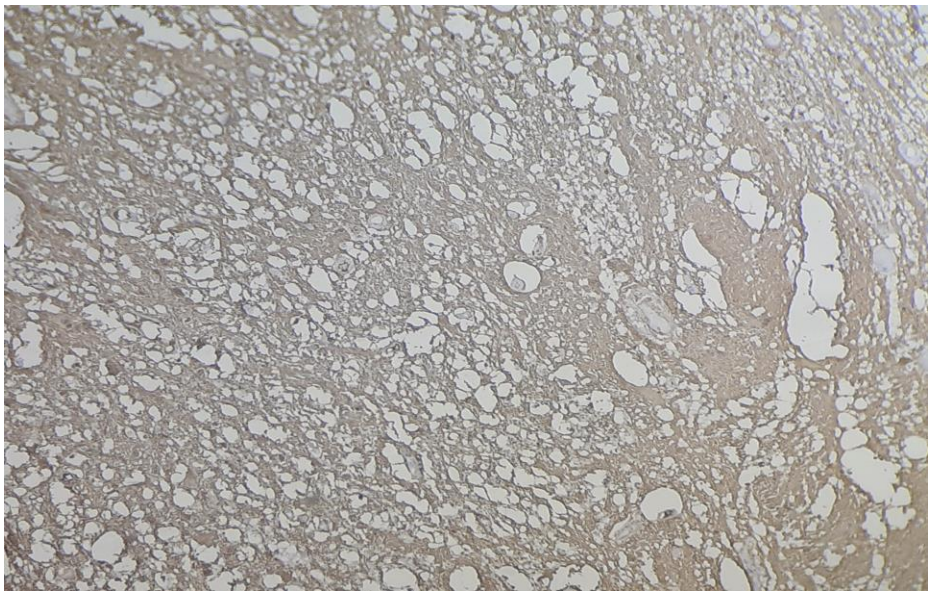

Figure 3- Tumors cells are EMA Positive

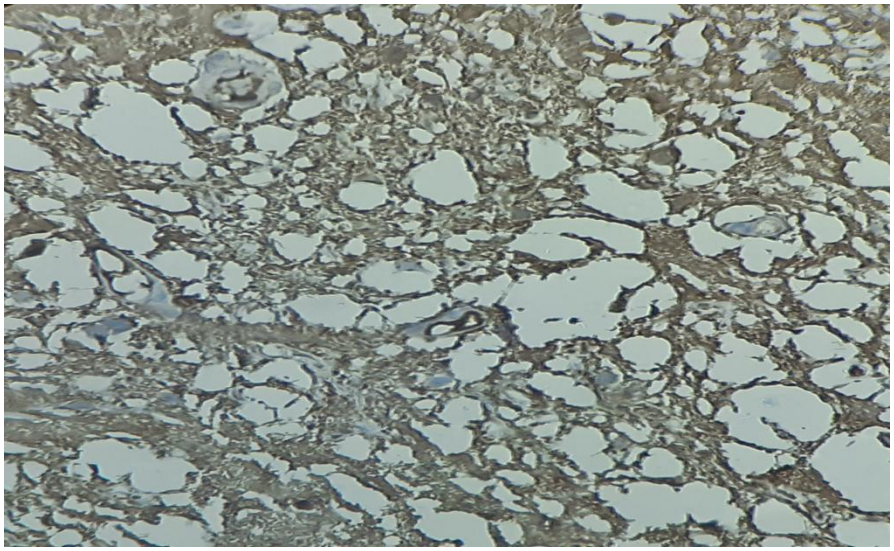

Figure 4- Tumors cells are S100 Positive

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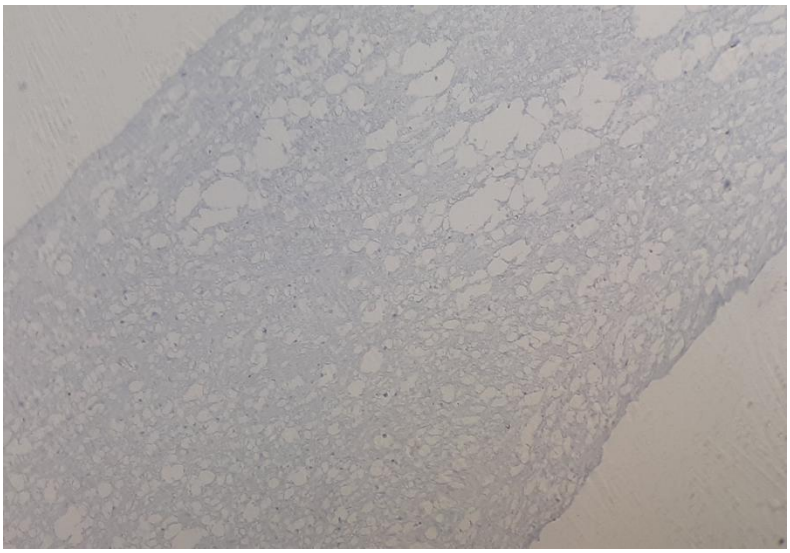

Figure 5- Tumors cells are CK7 Negative

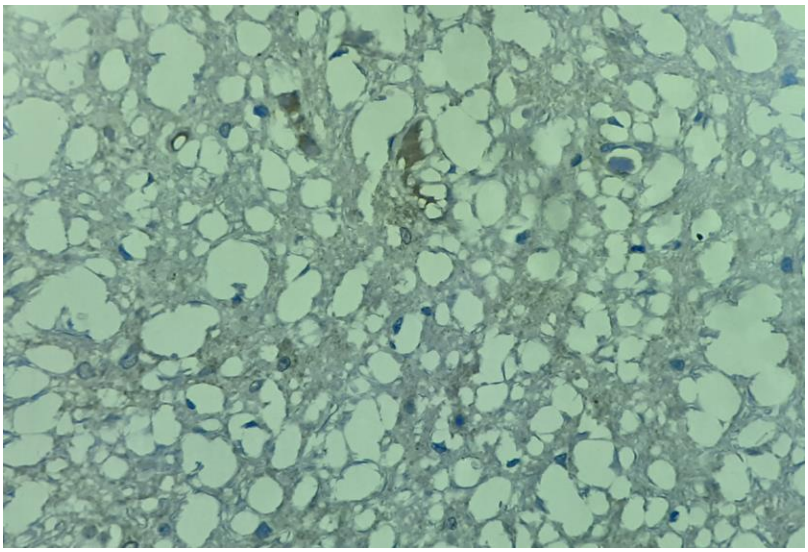

Figure 6- Tumors cells are CK19 focal positive