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Two rare localizations of chondrosarcoma: cervical and thoracic spine

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ABSTRACT

Chondrosarcoma is a primary malignant bone tumor, with spinal involvement being rare. Diagnosing this condition can be challenging and typically requires histological verification. Chondrosarcoma has the potential to invade locally and spread systemically.

We report the case of 32 year-old women presenting with back pain and weight loss. Magnetic resonance imaging of the spine showed a bone growth at the low cervical spine which extends to the upper dorsal spine. Histological examination showed low-grade chondrosarcoma. Given the tumor proximity to the spinal cord, an incomplete surgical excision was performed followed by radiotherapy sessions. Clinical course was marked by partial regression of the tumor process with a stability of the radiological image of the tumor after a three years follow-up.

Our case is original because of the bifocality of the chondrosarcoma in two adjacent segments of the spine.

Keywords: chondrosarcoma, cervical spine, dorsal spine

INTRODUCTION

Chondrosarcoma is a malignant tumor characterized by the proliferation of cartilaginous tumor cells. Unlike other bone tumors, it does not produce an osteoid bone component, which distinguishes it from osteosarcoma and other osteogenic tumors [1]. This tumor type is relatively rare in the vertebral bodies, with occurrences ranging from 2% to 12% of all cases [2]. When it does affect the spine, involvement of the cervical and thoracic regions is even less frequent compared to the lumbar spine, where the tumor is more commonly found.

The treatment for chondrosarcoma primarily focuses on surgical intervention. Complete surgical resection is critical, as it is the most effective way to achieve local control of the tumor and improve overall prognosis. However, the surgical procedure is often complicated by the tumor's invasive nature and its location near vital anatomical structures, such as the spinal cord and major blood vessels. The proximity to these critical structures can make complete excision difficult and increase the risk of surgical complications.

Additionally, the rarity and complex presentation of chondrosarcoma in the spine necessitate a multidisciplinary approach to treatment, often involving orthopedic surgeons, neurosurgeons, and oncologists. Preoperative imaging and careful surgical planning are essential to navigate the tumor's invasiveness and proximity to critical structures, while postoperative follow-up is crucial to monitor for any potential recurrence or complications.

OBSERVATION

A 46-year-old woman was admitted in our internal medicine department for weight loss of 20 kg in two months, inflammatory back pain and inter-costal neuralgia. Symptoms started two years prior to her hospitalization. Physical examination revealed a fixed, painless swelling adjacent to C7-T1, with no local inflammation signs. Neurological examination showed deep tendon hyper-reflexia on both lower

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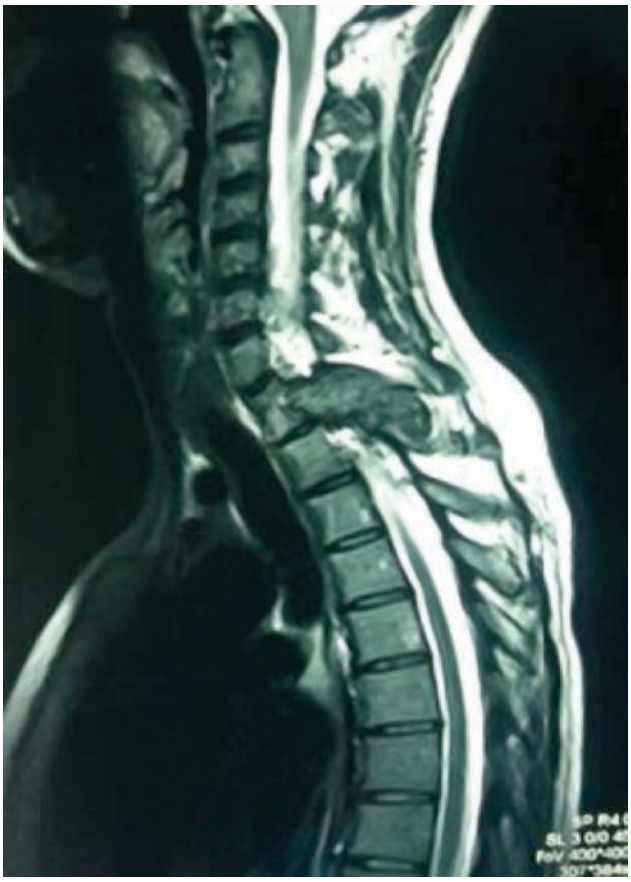

FIGURE 1. Sagittal plane MRI showing bone growth developing from the transverse processes of T1 and T2

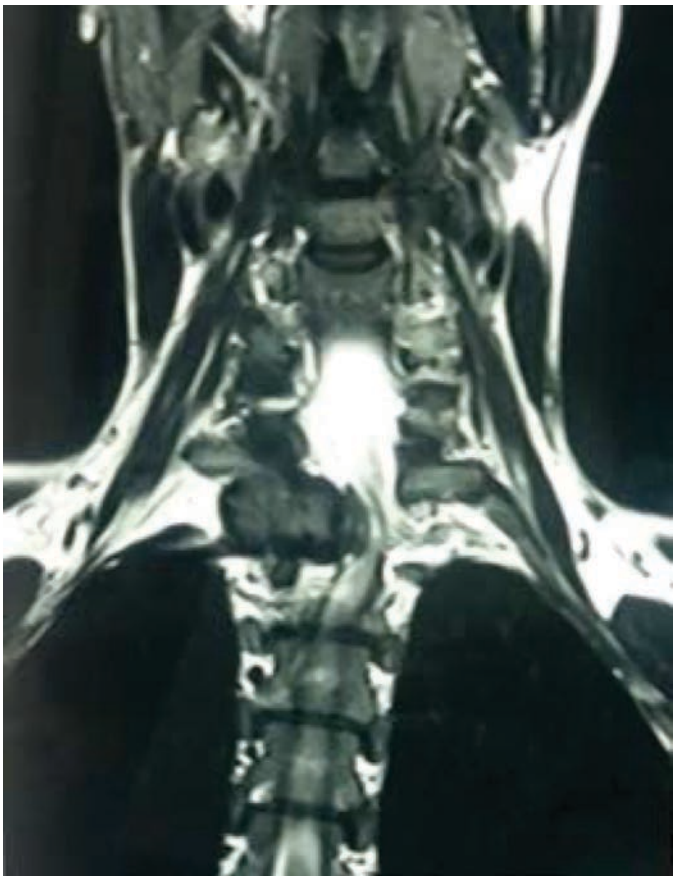

FIGURE 2. Coronal plane MRI showing spinal cord deviation between the pedicles by tumor process

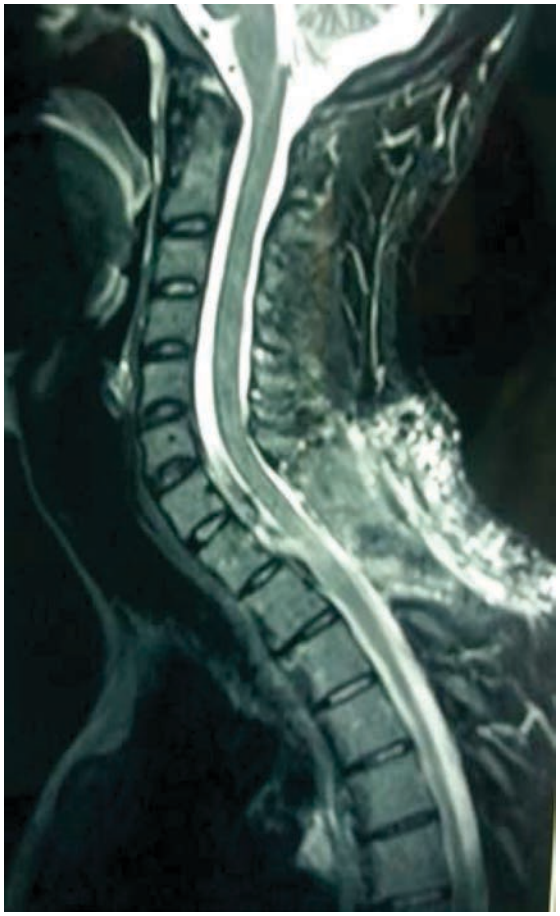

FIGURE 3. Sagittal plane MRI showing regression of tumor

limbs with a C4 sensory level. There was no laboratory evidence of inflammation, C-reactive protein level was 6 mg/dL and erythrocyte sedimentation rate was 32 mmH1. Full blood count was normal. Phosphate-calcium balance and liver function tests were within normal range. Magnetic resonance imaging of the spine showed a bone growth developing from the transverse processes of T1 and T2. Spinal cord was deviated between the pedicles from C7 to T3. Abnormality signal T2 of the spinal cord C7 toT2 was noted (Figures 1 and 2).

Partial surgical excision of the tumoral processes was performed. The tumor was rock solid with little spotting and spongy texture.

Histological examination showed tumor cells with cartilaginous differentiation destroying and infiltrating the spongy bone structures. It was composed of polygonal and globular chondrocytes and there was a cartilaginous structure in the form of lobules. Cytoplasmic volume was variable with atypical and irregular nucleus and the nucleolus was visible. Calcium deposits and tumor necrosis were found. These aspects were highly concordant with the diagnosis of a well differentiated, low grade Chondrosarcoma.

The patient had to undergo radiotherapy sessions since surgical excision was incomplete. After two months of surgical treatment and radiotherapy,

patient presented clinical improvement with clear decrease of back pain and a weight gain of 3 kg. MRI control showed a regression of the processes (Figure 3).

DISCUSSION

Chondrosarcoma ranks as the third most common primary malignant bone tumor, coming after osteosarcoma and Ewing sarcoma in terms of prevalence [1]. Although chondrosarcoma can occur in various locations within the body, its incidence in the spinal column is relatively rare, estimated to be between 2% and 12% of all chondrosarcoma cases [2]. When chondrosarcoma does affect the spine, it predominantly involves the lumbar region, which accounts for approximately 68% of cases. The thoracic spine is affected in about 23% of cases, while involvement of the cervical spine is rare, occurring in only 9% of cases [2].

Patients with chondrosarcoma typically present with dull, aching pain that is often exacerbated at night. This type of pain can be persistent and may interfere with daily activities and sleep, contributing to a reduced quality of life. The pain associated with chondrosarcoma is usually a result of the tumor's growth and its impact on surrounding tissues, including nerves and other structures within the spinal column. As the tumor progresses, it can cause additional symptoms such as neurological deficits, weakness, or changes in sensation, depending on its location and extent of involvement.

In our case, the pain was not severe but was characterized by significant radiation. The physical examination findings in chondrosarcoma patients vary based on the lesion's location. Typically, chondrosarcoma arises in the posterior part of the vertebrae. In 40% of cases, it remains localized, while in 45% it extends to the vertebral body. In some instances (15%), the tumor may be limited to the vertebral body alone [3]. Our case was original because chondrosarcoma was localized at both the low cervical spine and upper dorsal spine. The importance of the tumor mass and its local invasion affecting the spinal cord contrasts with minor clinical signs and can be attributed to slow evolution.

Diagnosis of chondrosarcoma was confirmed by histological examination. It showed tumor lobules of irregular size and shape delimited by connecting septa. Residual spongy bone fragments sometimes persisted within these lobules. The tumor cells were bi-nucleated with minimal to moderate atypia; poorly limited forming lobules with abundant cartilaginous matrix, separated by narrow fibrous bays. The matrix was solid or myxoid with presence of calcifications and bone metaplasia [4].Tumor cells looked like chondroma. In fact, a major problem of cartilaginous tumors is the distinction between benignity (chondroma, chondromatosis) and malignancy (low grade chondrosarcoma) [5].

In fact, the distinction between benign and malignant often depends on evidence of a “chondrosarcoma penetration pattern,” in which the tumor infiltrates through the medullary cavity rather than being confined to the native structure, which is associated with poor prognosis [6,7].

Grade I chondrosarcoma is characterized by moderate cellularity and features hyperchromatic nuclei that are uniformly sized and thick. In contrast, Grade II chondrosarcomas exhibit increased hyperchromasia, greater nuclear atypia, and larger nuclear sizes, along with a higher degree of cellularity. Mitoses may be present in these tumors.

Grade III chondrosarcomas show even greater pleomorphism and atypical features compared to Grade II tumors. They are more cellular, with mitoses being more readily identifiable. Cells at the periphery of these tumors often appear less differentiated and adopt a spindle-like shape.

According to recent literature, the histological grading of chondrosarcoma plays a crucial role in determining prognosis and guiding treatment strategies. Studies have shown that higher-grade tumors, such as Grade III, are associated with a more aggressive clinical course and a higher likelihood of metastasis, necessitating more intensive treatment approaches and closer follow-up

The fourth category of chondrosarcoma, known as Grade IV, is referred to as dedifferentiated chondrosarcoma. This subtype accounts for approximately 10% of all chondrosarcoma cases. Dedifferentiated chondrosarcoma is characterized by a high-grade tumor that often appears fusiform or pleomorphic and lacks significant cartilage matrix. Histologically, it is defined by the presence of both a low-grade cartilaginous component and a high-grade non-cartilaginous component, which may be more aggressive and poorly differentiated.

According to recent literature, dedifferentiated chondrosarcoma is considered a high-grade tumor with a notably poor prognosis due to its aggressive nature and higher likelihood of metastasis. The prognosis for patients with this subtype is generally worse compared to other chondrosarcoma grades, and it often requires more aggressive treatment approaches and closer monitoring.

In addition to dedifferentiated chondrosarcoma, other rarer forms include mesenchymal chondrosarcoma and clear cell chondrosarcoma. Mesenchymal chondrosarcoma is distinguished by its rapid growth and high metastatic potential, whereas clear cell chondrosarcoma is characterized by a distinct histological appearance with clear cells and a more variable clinical course.

Chondrosarcoma can lead to several complications, including the risk of tumor recurrence and distant metastases, predominantly affecting the lungs. The likelihood of metastasis varies significantly based on the tumor grade. For low-grade chondrosarcomas, the recurrence rate is typically below 10%. In contrast, moderate-grade chondrosarcomas have a recurrence rate ranging from 10% to 50%. High-grade chondrosarcomas present a much greater risk, with metastasis occurring in approximately 50% to 70% of cases.

These variations underscore the critical need for stratifying patients based on tumor grade and stage to accurately evaluate their prognosis and tailor treatment strategies accordingly. Effective management and close monitoring are essential to address potential complications and optimize patient outcomes. Recent studies emphasize that careful assessment and proactive intervention can significantly impact the management of chondrosarcoma and improve overall prognosis.

The primary treatment for chondrosarcoma is surgical excision. Achieving complete excision is a significant predictor of improved survival outcomes, both for tumors that are confined or locally invasive and for those that have metastasized. However, the invasive nature of chondrosarcoma and its proximity to critical structures such as the central nervous system and the aorta can make surgical resection particularly challenging.

For cases where complete surgical resection is not feasible, such as in advanced disease or subtotal resection cases like that of our patient, adjuvant radiotherapy is often employed to address residual tumor and reduce the risk of recurrence.

Chondrosarcoma generally exhibits resistance to chemotherapy and immunotherapy, making these treatments largely ineffective for conventional forms of the disease [3]. Nonetheless, chemotherapy may be considered in cases of dedifferentiated chondrosarcoma that include a high-grade spindle cell component, where it can sometimes offer therapeutic benefits. Recent studies emphasize the limited efficacy of standard chemotherapeutic approaches for conventional chondrosarcoma, highlighting the need for alternative or adjunctive treatment strategies in managing this malignancy.

The prognosis for chondrosarcoma located in the axial skeleton generally appears to be less favorable compared to chondrosarcoma affecting the long bones. For long bone chondrosarcomas, the 5-year survival rate is estimated to range from 50% to 80% [6, 8-10]. Several factors influence the prognosis of chondrosarcoma, including histological differentiation [8,9], age at diagnosis, tumor size, race (with Black patients showing a potentially higher risk), tumor grade, and the extent of disease [2].

In the case under discussion, the patient did not require assistance from a care manager. Instead, family members provided crucial support, including transportation and assistance during medical appointments. This support system is essential in managing the logistical and emotional aspects of the disease, highlighting the role of family involvement in patient care and treatment adherence.

CONCLUSION

Chondrosarcoma occurring in the cervical and thoracic spine is extremely rare. Diagnosing this condition can be challenging due to the nonspecific nature of its symptoms and the histological similarities between chondroma and low-grade chondrosarcoma. These similarities often complicate the differentiation between benign and malignant forms of the tumor.

Surgical excision is generally considered the preferred treatment approach for chondrosarcoma, as it offers the best chance for removing the tumor and improving patient outcomes. The prognosis for chondrosarcoma is often poor, primarily due to its potential for local invasion, which can lead to the destruction of surrounding structures, and its tendency to metastasize to distant sites. These factors contribute to the overall complexity and seriousness of managing this malignancy, underscoring the need for early and accurate diagnosis, as well as effective surgical intervention to improve survival rates and minimize the risk of metastasis.

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The authors undersign and certificate that they do not have any financial or personal relationships that might bias the content of this work.

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