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Case Report

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Lumbar Chondrosarcoma in A Young Female: A Case Report and Literature Review

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ABSTRACT

Introduction: Chondrosarcomas (CS) are uncommon malignant tumors that form cartilage, ranking third among primary malignant bone tumors. Although they are rare, with an incidence rate of 2%-12%, they can occur in the spine, with the lumbar region being even less common. These tumors are resistant to chemotherapy and radiotherapy, making surgical excision the treatment of choice. The extent of surgical resection is vital for the patient's prognosis and the risk of recurrence. A unique case of lumbar CS in a young female patient is reported.

Case Report: A 23-year-old female experienced progressive lower back pain and radiculopathy in both lower limbs. Laboratory tests and radiological imaging revealed an osteolytic lesion in the L5 vertebral body. Despite a biopsy that did not conclusively diagnose the lesion, a two-stage en-bloc resection was carried out. The procedure confirmed the diagnosis of grade I conventional chondrosarcoma. The patient had a successful recovery and remained recurrence-free after three years.

Discussion: CS typically occurs in males between their 40s and 70s and can develop in various bones, with different classifications based on origin and histopathological features. Spinal CS is rare, and symptoms can be non-specific but may progress to include pain and neurological deficits. Diagnosis is primarily based on imaging, with biopsy used in some cases. Surgical resection is the mainstay of treatment due to the tumor's resistance to other treatments. Achieving negative surgical margins is imperative but challenging, necessitating a multidisciplinary approach for optimal outcomes.

Conclusion: The presented case underlines the rarity of CS in the lumbar region and the challenges in diagnosing and surgically managing such cases. Aggressive surgical resection is key to treatment, requiring careful preoperative planning and intraoperative precision. The successful long-term disease control in our patient emphasizes the importance of complete tumor resection.

Keywords: Lumbar Spine, Chondrosarcoma, Sarcoma, Soft Tissue Tumor, Spondylectomy

Introduction

Chondrosarcomas (CS) are heterogenous group of malignant tumors originating from chondroid (Cartilaginous) matrix with an ability of cartilage formation [1]. Although it is the third most common primary malignant tumor, it is considered rare with an estimated incidence of 1 in 200,000 per year [2]. CS can be categorized into primary or secondary depending on their origin; de novo or from preexisting cartilaginous lesion such as osteochondroma [3]. It can originate in different locations where central CS arise in intramedullary cavity of bone, while the peripheral CS arise on the cortical bone secondary to preexisting lesion [4]. There are variety of subtypes with different histopathological features and clinical behavior [1]. Conventional chondrosarcoma counts for most cases and comprise around 75% to 85% of low to intermediate grade tumors. Other less common variant types include myxoid chondrosarcoma, mesenchymal chondrosarcoma, dedifferentiated chondrosarcoma, and clear cell chondrosarcoma. [5,6] It usually arises in the trunk or proximal femur and humeri with a being the place that is less commonly involved [1,2].

Spine considered the primary location in only 2% to 12% of chondrosarcoma cases [1,4,7]. It can affect any segment of the

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spine with thoracic spine being the most affected followed by the cervical and lumbar region with scattered cases of sacral and coccyx involvement [3,6-8]. Tumors may develop from any one of the vertebra's three primary growth zones with approximately 5% of these tumors emerge from the vertebral body, 40% from the posterior elements, and 45% involve both areas [6]. Chondrosarcomas are most commonly identified in individuals aged between 30 and 70, with a marginally higher prevalence observed in males compared to females [6,8]. Pain is usually the presenting symptom, with mass in the involved region and neurological symptoms being commonly reported [2,9,10].

CS are known to be resistant to the typical treatment protocols of chemotherapy and radiotherapy; therefore, surgical excision must be attempted [2,3,6]. It is advocated that surgical intervention is the primary mode of treatment. Moreover, it has been noted that the extent of surgical resection has a pivotal role in the prognosis and recurrence [2]. Only few series on spinal CS were reported in the literature [3,6-8,10-16].

Herein, we present a case of young female who presented with progressive lower back pain that was associated with initial right radiculopathy that evolve to involve both lower limbs and found to have lumbar Chondrosarcomas.

Case Report

A 23-year-old female patient with increasing lower back pain for the past three months. The pain progressed to right lower limb radiculopathy and numbness then involve the left lower limb as well. Pain was noted to be aggravated by movement and mobilization. On examination, Power was 5/5 all over including hip flexion, hip adduction, knee flexion and extension, planter flexion, extensor hallucis longus and dorsiflexion. Knee reflexes were +2 and symmetrical bilaterally. Decrease sensation over right medial side of the leg and over the medial malleus (Right L4 dermatome) was evident as well. Positive straight leg raise test at 60 degrees in right lower limb. Upon admission, routine blood work up was done including CBC, electrolytes, coagulation profile and all were within normal ranges. Additionally, Inflammatory markers were obtained and revealed slightly elevated levels; ESR 23 mm/hour (Normal range 0-20) and CRP 8.38 mg/L (Normal range 1-3). Moreover, radiological studies showed osteolytic lesion in L5 vertebral body with well-defined margin with moderate retropulsion of posterior wall and cortical thinning and break through. In addition, there was mild soft tissue component causing severe spinal canal stenosis. (Figure1) MRI spine shows well defined lesion involving L5 vertebral body with geographic sclerotic margin measures 3 x 3.2 x 2 cm on AP, TR and CC dimensions with ring enhancement in T1 with contrast. (Figure2) Malignancy work-up and CT chest, abdomen and pelvis were done, and it was unremarkable apart from that L5 lesion.

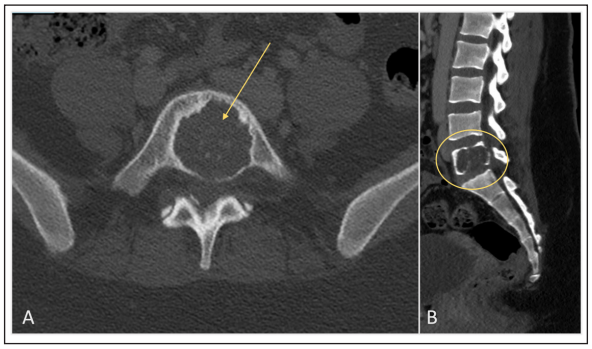

Figure 1: Preoperative images. Pre-op Computed tomography (CT) (A) axial and (B) sagittal showing osteolytic lesion in L5 vertebral body (yellow circle and arrow) with well-defined margin with moderate retropulsion of posterior wall and cortical thinning and break through. In addition, there was mild soft tissue component causing sever spinal canal stenosis.

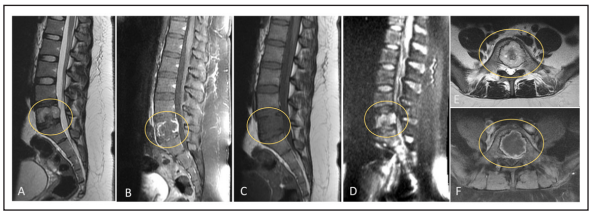

Figure 2: Preoperative images. Pre-operative magnetic resonance image MRI (A) sagittal T2, (B) T1 with contrast, (C) plain T1, (D) diffusion-weighted, (E) axial T2, (F) axial contrasted T1 showing a well defined lesion involving L5 vertebral body (yellow circle) with geographic sclerotic margin measuring 3x3.2x2 cm on AP, TR and CC dimensions with ring enhancement in T1 with contrast.

Due to the uncertainty of the lesion nature, initial management was CT guided biopsy from L5 vertebral body lesion which was insignificant. (Figure3)

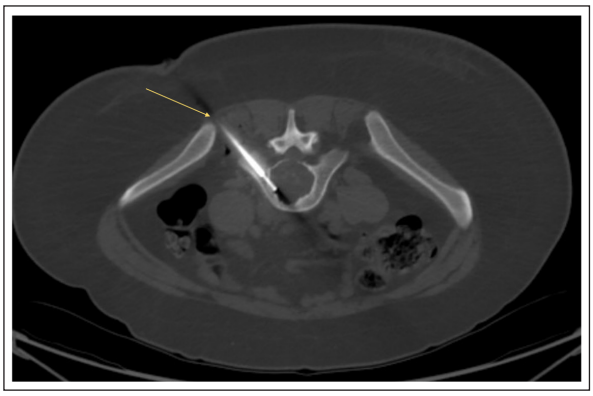

Figure 3: CT-guided biopsy approach from L5 vertebral body lesion (Yellow arrow).

Management options were discussed, and the patient subsequently underwent en-bloc resection with a goal of decreasing recurrence rate. Patient was operated on 2 stages; first one, was L5 laminectomy and Lumbar 3- Sacral 2 fixation and fusion through posterior approach. The second stage was the anterior lumbar approach for L5 corpectomy and cage insertion (Figure4) (Figure5).

The histopathological examination established a diagnosis of grade I conventional chondrosarcoma. (Figure6) Postoperatively,

the patient recovery was uneventful, and she showed progressive improvement with no neurological deficit. Patient was followed in the clinic for 2 years with no evidence of local recurrence. (Figure7).

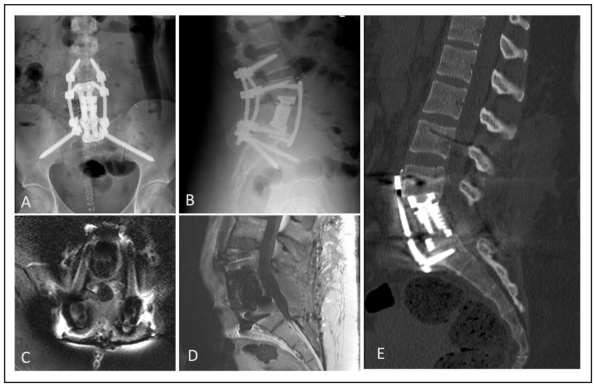

Figure 4: Post-operative images. (A) coronal x-ray, (B) sagittal x-ray, (C) axial contrasted T1 MRI, (D) sagittal T1 contrasted MRI and (E) sagittal CT showing satisfactory fixation and lesion resection.

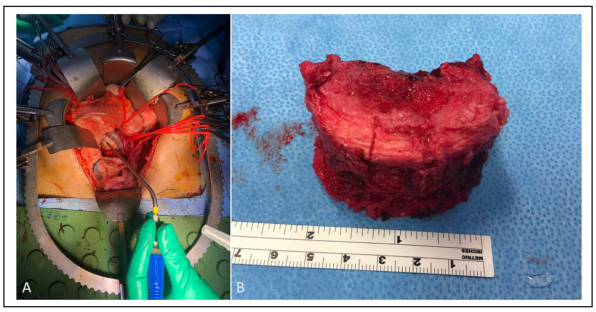

Figure 5: Surgical approach. Anterior approach (A) and gross picture (B) for en-bloc resection inform of L5 corpectomy.

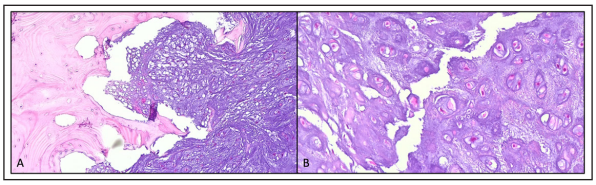

Figure 6: Histopathology. Hematoxylin and Eosin stain (A) shows chondrosarcoma with permeation of intertrabecular space (x10). (B) shows chondrosarcoma with minimally increased cellularity, nodular growth and occasional binucleate nuclei (x40).

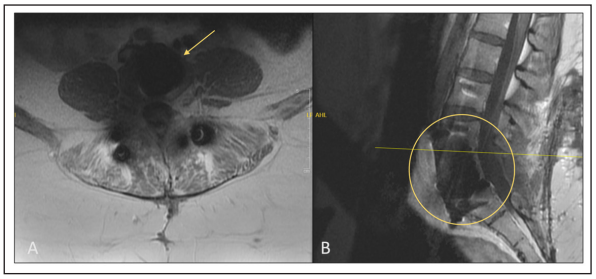

Figure 7: Follow-up. Latest follow up MRI (A) axial and (B) sagittal 3 years post-op showing no evidence of any local recurrence (yellow circle and arrow).

Discussion

CS constitutes a heterogenous group of bone tumors that affect the cartilage tissue [2]. It is a frequent bone malignancy that has been reported with variable incidence rate of 0.5 per 100,000 patient-years [2,9]. CS is generally accepted to be the third most common bone malignancy after osteosarcoma and Ewing's sarcoma [9]. It has a male predominance with a male: female ratio of 1.5:1 [1,9,17]. Also, it primarily affects patients between the 4th and 7th decade [4,8]. CS can be classified by origin as primary or secondary [9]. Primary tumors are the tumors arising in de novo also known as conventional chondrosarcoma which accounts for 85% of all cases. Secondary CS referred to the tumors that develop from the cartilage cap of sporadic osteochondroma or sporadic enchondroma [2]. Moreover, secondary chondrosarcoma can arise in patients affected by genetic syndromes like hereditary multiple exostosis, Ollier disease, or Maffucci syndrome [9]. Based on the location, CS can be classified as either central or peripheral within the parent bone. CS that arises within the intramedullary cavity are known as Central chondrosarcomas, whereas CS that arise on the surface of cortical bone are known as peripheral chondrosarcomas [1]. According to the histological features, conventional CS represent the most common type [8]. Other types include dedifferentiated, mesenchymal, juxtacortical, and clear cells [2,8,9].

Additionally, CS can involve any skeletal bone; however, it tends to commonly affect long tubular bones, pelvic bone, shoulder girdle, and ribs [3]. Spine is a location that is less commonly involved with a reported rate of 2% to 12% of cases. Among spine cases, thoracic spine is the most favorable location followed by lumbar, cervical, and lastly sacral cases [3,6]. Lesions may arise in either the vertebral body (5%), the posterior elements (40%), or both (45%); since there are three growth centers in each vertebra from which the tumor originates [6].

CS has variable clinical presentations depending on the size, location, and grade of the tumor [9]. Symptoms can be non-specific, or patient can present with pain, mass, and tenderness as tumor grows bigger. Also, patients can present with neurological symptoms such as radiculopathy and myelopathy if the tumor is involving the spinal column [9].

Radiologically, CS typically demonstrate "ring and arc" pattern which results from chondroid matrix mineralization [6,9]. Magnetic resonance imaging (MRI) and computed tomography (CT) are diagnostic tools that are frequently utilized to evaluate the tumor extent, determine the stage, and plan the surgical resection [9]. Furthermore, radiological advancement can be utilized to obtain a biopsy as core needle biopsy or fine needle aspiration can be helpful in yielding diagnostic information to determine the next step in management [9]. Pennington et, al has suggested an algorithm for chondrosarcoma management [9]. Surgery remains the mainstay for CS management as they are relatively resistant to radiotherapy and chemotherapy due to slow growth of tumor and poor vascularization [6]. Achieving complete resection with negative margins when surgically treating lumbar CS presents several challenges. The complexity of the surgical procedure and the associated risks are influenced by numerous factors, including the tumor's anatomical location, size, biological behavior, and involvement of critical structures. Treating team should aim for complete resection as it can be curative in low grade tumors and en-bloc resection with tumor free margin can decrease the rate of recurrence in low-grade tumor and can help to prevent metastasis in high-grade tumors [9,18].

Although prognosis depends on multiple variables like histologic grade, stage, and location of the tumor, surgery is the only modifiable factor that can improve prognosis [6,9,16]. In general, low-grade CS has a good prognosis with 10-year survival ranging from 89% to 95% [18]. Few studies have discussed surgical approaches for achieving en-bloc resection in lumbar CS [3,16,19,20].

Here we presented a rare case of young female with diagnosis of lumbar CS with no recurrence for more than 3 years. In this case, although initial biopsy was not inclusive, surgical biopsy yield a diagnosis that shaped the surgical management as patient was operated on two-stages to achieve complete resection and stability. This case aims to emphasize the importance of complete en-bloc resection despite complex structure and demanding technique and highlight surgical technique used to achieve best possible outcome.

Conclusion

Primary CS of the lumbar vertebrae are extremely rare. Tumor in the spine represents a challenge due to its complex nature and anatomical constrictions. Despite limited clinical evidence, surgery remains the gold standard in management of CS cases and total en-bloc resection should be considered. Surgeons must carefully balance the goal of negative margins with the risk of morbidity. Preoperative planning with detailed imaging helps in understanding the tumor's relationship with surrounding structures. Intraoperative navigation and monitoring may also help in achieving a safer resection. Multidisciplinary teams should collaborate to optimize the surgical approach and improve outcomes for patients with lumbar CS.

Authors Contribution: All authors contributed to the study conception and design. Data collection and manuscript writing were performed by Moudi AlHussainan, Khulood K. AlRaddadi and Abdulealah AlTurkistani. Figures were prepared by Saad AlObaisi and Khulood K. AlRaddadi and Salman T. Almalki. Supervision by Abdullah Labeeb. All authors read and approved the final manuscript. All authors have approved the manuscript and agree to its submission to this journal.

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