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Orthopaedics

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SECONDARY CHONDROSARCOMA OF LUMBAR SPINE- A CASE REPORT

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ABSTRACT

Introduction: Chondrosarcoma is a malignant cartilage forming bone neoplasm that accounts for 10% of all primary bone tumors. Most occur in the thoracic spine followed by lumbar spine and patients typically present in middle age with back pain and/ or neurological symptoms. Radiologically these tumors appear as destructive lesion in the spine or as a paraspinal mass with calcification. Malignant degeneration is the most serious complication, however it is rare and found in 5% of cases of multiple hereditary exostosis(1).We describe a case of man in his early 50s who presented with features and diagnosed to have chondrosarcoma of lumbar spine. **Case Report**: 54 years old male came with chief complaints of low back pain for 1.5 months with complaints of weakness over both lower limbs - 25 days, with bowel and bladder dysfunction, history of loss of weight, loss of appetite. No other comorbidity and no relevant family history found. MRI lumbar spine shows large exophytic lesions with irregular sclerotic foci from posterior elements of L4-L5 vertebra with soft tissue component. Tumour excision and biopsy confirms that patient had an uncommon diagnosis of Chondrosarcoma of lumbar spine for which he was managed accordingly. **Conclusion:** Early diagnosis and complete resection of tumor and treatment of relevant symptoms represent a viable treatment for this rare disorder to achieve increased life expectancy, low recurrence of tumor, and improvement of quality of life.

INTRODUCTION:

Chondrosarcoma of spine is a heterogeneous group of Cartilage forming tumors. It is the third most common Primary malignant bone tumor. [1] [2] Incidence is 2% to 12% to various sites. [2]

Most common site is the thoracic Spine followed by the cervical & lumbar region. Most frequent location were vertebral body and posterior elements (45%). Posterior elements (40%) and vertebral body (5%). [2]

Chondrosarcoma of the spine may arise denovo or from the pre-existing cartilagenous lesion such as an osteochondroma or enchondroma. Presenting symptoms includes pain in most of the cases, and the other complaints were palpable mass & neurological deficits.

The ideal & most successful treatment for the Spinal chondrosarcoma is complete enbloc resection of the tumor. Our case report is on a Chondrosarcoma of lumbar spine in a patient with multiple hereditary exostosis at multiple sites, which is less common.

Case Report:

54 years old male presented with chief complaints of low back pain for 1.5 months with increase in size of swelling which was present since childhood for past 1 month and complaints of weakness over both lower limbs - 25 days, weakness was present initially in the left lower limb then it involved right lower limb also, bowel and bladder incontinence, not able to ambulate for past 5 days. Patient is a known case of multiple hereditary exostosis with multiple bony swelling since childhood. Patient had history of loss of weight, loss of appetite.

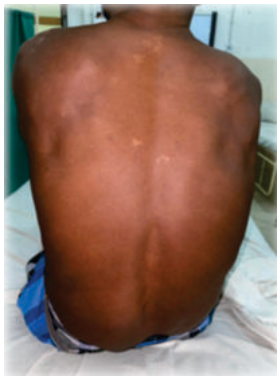


Figure: 1. Preop Clinical Image Of Patient

No other comorbidity and no relevant family history found.

Local Examination of Lumbar spine:

Inspection

Swelling present over left paraspinal lumbar region in L2-L4, ovoid swelling of size 15x8x3cm over lumbar region in L2-L4 Left side, List present, No kyphosis.

On Palpation tenderness present in the left paraspinal lumbar

region L2-L5 which is firm in consistency, immobile with irregular surface extending from L2-L5. Skin over the swelling mobile

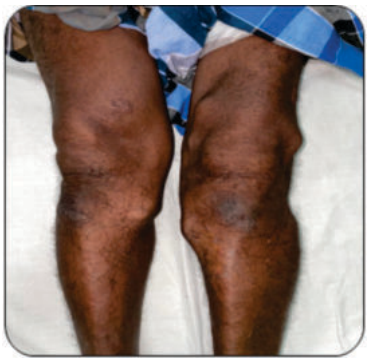


Figure 2: Preop Gross Clinical Image

Neurology:
Motor neurology (L2) iliopsoas on both sides found to be 3/5 on both sides. Quadriceps L4 (4/5 on both sides). Tibialis anterior, Extensor hallucis longus & tibialis posterior were found to be 2/5 & 1/5 on left side.

Sensory from L3- S1 dermatome is reduced on both sides spanning L2 dermatome with diminished knee and ankle reflex, with reduced perianal sensation with lax anal tone with grade I bed sore over left gluteal region.

Investigations:
Patients haemoglobin was found to be 14.4 g/dl with total count 12 26 x 1000/ microliter. C reactive protein was 19 mg (positive for C reactive protein with ESR was 7mm/hour with elevated alkaline phosphatase 137 U/L.

CT lumbar spine shows large exophytic lesions with irregular sclerotic foci from posterior elements of L4-L5 vertebra with soft tissue component in the left para spinal region with apparent extension at L3, L4 levels to spinal canal and left neural foramina.

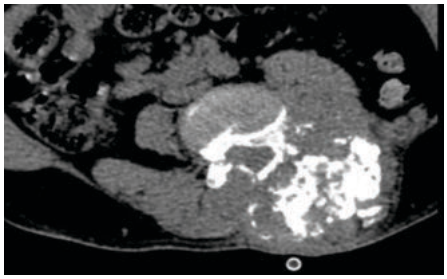


Figure 3: Large exophytic lesion with irregular sclerotic foci from osterior elements of 14, 15 vertebra with soft tissue component

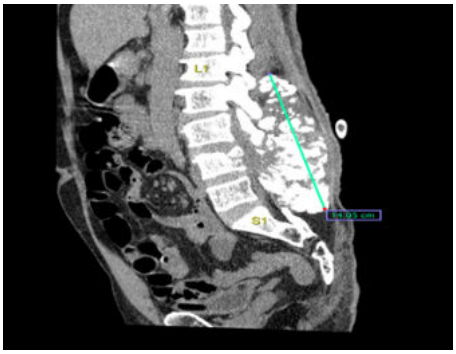


Figure 4: CT SPINE-Large exophytic lesions with irregular sclerotic foci from posterior elements of L4,L5 vertebra with soft tissue component in the Left para spinal region with apparent extension at L3,L4 levels in spinal canal and left neural foramina

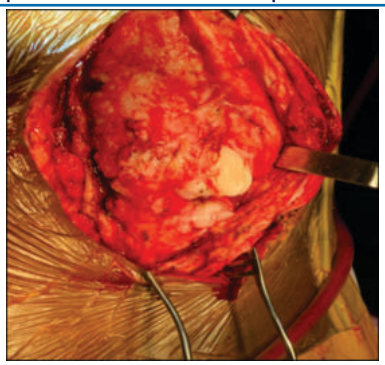


Figure 5: Intraop Gross Picture Of Excision And Biopsy

Treatment:
- Patient underwent Tumour excision biopsy with decompression and stabilisation from L3 to L5.
- PROCEDURE: Chondrosarcoma excision under epidural and general anaesthesia Patient in prone position, parts painted and drapped.
- Through curved incision on left side L3-S1, lamina exposed, tumour margins delineated mediolaterally and superoinferiorly.
- Medially tumour was found extending upto L4 lamina on right side.
- L3-L4 spinous process osteotomized and tumour deliverd out.
- 18x 8x 6 cm dimensions of tumour.
- Tumour tissue was found to be encroaching under L3-L4 posterior elements.
- Decompression done and tumour tissue dissected off the dura and removed.
- Frank pus noted in left psoas region and sent for culture and sensitivity. Right L3,L4,L5 pedicles instrumented with polyaxial titanium pedicle screws. Rod connected. Wound washed, hemostasis achieved, wound closed in layers and drain insitu.

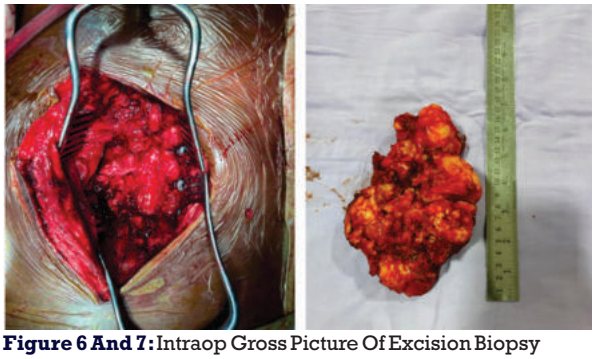


Figure 6 And 7: Intraop Gross Picture Of Excision Biopsy

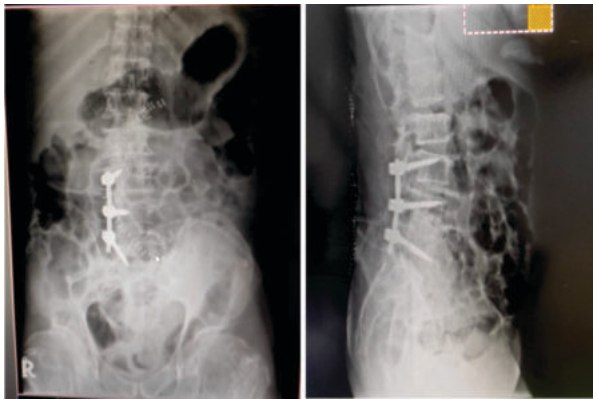


Figure 8: Post Stabilisation Xray Of Lumbar Spine

Post operatively patient was hospitalised for 14 days and was started on INJ.VANCOMYCIN and INJ.MEROPENAM. On POD-12 suture removal was done and patient has been referred to

regional cancer institute and Patient currently on chemotherapy 6 cycles of Ifosfamide, Adriamycin and opioid derivatives for pain management. Patient was on regular follow up and he is symptomatically better.

DISCUSSION:

Chondrosarcoma developmental lesion of the bone is a result of ectopic development of growth plate of cartilage. The epiphyseal cartilage separated from the normal growth plate herniated into the periosteal bone cuff. The enlargement of this cartilaginous fragment and its eventual enchondral ossification explained that excrescent growth of osteochondroma from bone surface and its cartilage cap.[3][4]

Hereditary Multiple Enchondromatosis is autosomal dominant inheritance pattern with incomplete penetrance in female. In HME patients genetic analysis detected abnormalities in three different loci of chromosomes in 8,11,19.[5][6]

They were termed EXT-1 on chromosome 8q 23-q24, EXT 2 on 11p- p12 and EXT 3 on short arm of chromosome 19. Inactivation of EXT gene causes osteochondroma formation and malignant transformation.[5][6] HME diagnosed before the age of 12, children older than 12 without the lesion detected will not manifest the condition.

Features Of Malignancy Includes

- The following growth of a previously unchanged osteochondroma and skeletally matured patients.
- Irregular or indistinct lesion surface
- Focal regions of radiolucency of the interior of the lesions Erosion or destruction of adjacent bones
- Significants of tissue mass particularly containing scattered or irregular calcification
- Hyaline cartilage cap thickness a reliable indicator of malignancy
- Cartilage cap thickening cutoff of 2cm
- MRI can have sensitivity and specificity of 100% and 98% respectively for detection of malignant transformation.

Secondary Chondrosarcoma

It arise most commonly from osteochondromas of multiple familial type.It's a chondrosarcoma arising from the cartilaginous cap part of the preexisting osteochondroma. Ollier disease is an isolated enchondromatosis with dedifferentiation incidence of 10-30% and Maffucci syndrome is enchondromatosis associated with soft tissue hemangiomas with dedifferentiation risk as high as 57%.[3][4] Our case is a case of Secondary dedifferentiated chondrosarcoma from primary multiple hereditary exostosis.

Grading:

Chondrosarcomas are graded based on cellularity and cytology. Grade I are moderately cellular with hyperchromatic nuclei of uniform size, Grade II will be hypercellular with moderate nuclear atypia. Grade III has pleomorphic, atypical cells and spindle cell. Our case was a Grade 3 dedifferentiated chondrosarcoma with intermittent areas of low grade component which looked tan or pale yellow and fleshy with a blue-grey cartilaginous area (low-grade component) with histology showing extreme cellularity with large bizarre nuclei and atypical cells.[8][9].

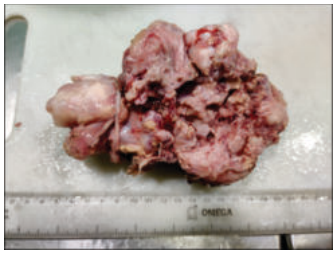
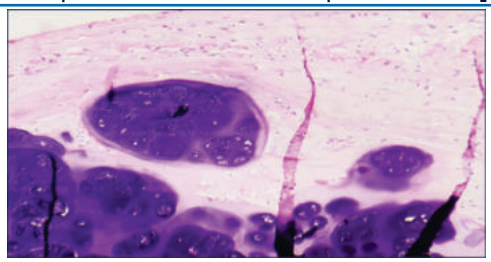


Figure 9: Postoperative Gross Pathological Specimen



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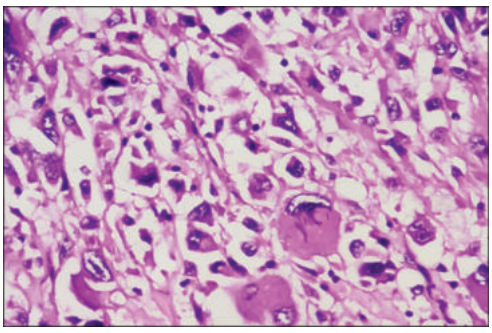

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Treatment Of Secondary Chondrosarcoma:

Grade I, Grade II and Grade III secondary chondrosaromas should be treated with total resection, which controls the local spread, recurrence from the free margin and disease specific survival. Chondrosarcomas has been found to be radiosensitive. However, stereotactic beam radiotherapy and proton beam radiotherapy are also found to be beneficial for the local control[6][7] and consequently long-term survival.

CONCLUSION:

In terms of clinical presentation and MRI findings chondrosarcoma spine is rare entity, surgical resection confers survival benefit in patient with chondrosarcoma of the spine independent of extent of disease. Radiotherapy improves survival in patients with metastatic disease and worsens outcome in patient with confined and locally invasive disease.

References:

1. Katonis P, Alpantaki K, Michail K, Lianoudakis S, Christoforakis Z, Tzanakakis G, Karantanas A. Spinal chondrosarcoma: a review. Sarcoma. 2011;2011:378957. doi: 10.1155/2011/378957. Epub 2011 Mar 8. PMID: 21437176; PMCID: PMC3061459.
2. Strike SA, McCarthy EF. Chondrosarcoma of the spine: a series of 16 cases and a review of the literature. Iowa Orthop J. 2011;31:154-9. PMID: 22096435; PMCID: PMC3215129.
3. Pennington Z, Ehresman J, Pittman PD, Ahmed AK, Lubelski D, McCarthy EF, Goodwin CR, Sciubba DM. Chondrosarcoma of the spine: a narrative review. Spine J. 2021 Dec;21(12):2078-2096. doi: 10.1016/j.spinee.2021.04.021. Epub 2021 May 8. PMID: 33971325.
4. Arockiaraj J, Venkatesh K, Amritanand R, Sundararaj GD, Nachimuthu G. Chondrosarcoma of the spinous process: a rare presentation. Asian Spine J. 2012 Dec; 6(4):279-83. doi: 10.4184/asj.2012.6.4.279. Epub 2012 Dec 14. PMID: 23275812; PMCID: PMC3530703.
5. Panelos J, Voulgaris S, Michos E, Doukas M, Charalabopoulos K, Batistatou A. Chondrosarcoma of the spine: a rare case with unusual presentation. Diagn Pathol. 2006 Oct 30;1:39. doi: 10.1186/1746-1596-1-39. PMID: 17074093; PMCID: PMC1634757.
6. Lin M, Zhou H, Zhang X, Hu Y, Guo T, Shi J. Primary mesenchymal chondrosarcoma of the adult lumbar spine: a case report and review of the literature. Transl Cancer Res. 2022 Sep;11(9):3363-3370. doi: 10.21037/tcr-22-122. PMID: 36237229; PMCID: PMC9552263.
7. Ha TH, Ha TMT, Nguyen Van M, et al. Hereditary multiple exostoses: A case report and literature review. SAGE Open Medical Case Reports. 2022;10. doi:10.1177/2050313X221103732.
8. Legeai-Mallet L, Munnich A, Maroteaux P, et al. Incomplete penetrance and expressivity skewing in hereditary multiple exostoses. *Clin Genet* 1997; 52(1):12–16. Crossref. PubMed.
9. Carroll KL, Yandow SM, Ward K, et al. Clinical correlation to genetic variations of hereditary multiple exostosis. *J Pediatr Orthop* 1999; 19(6): 785–791. Crossref. PubMed
10. Unni KK, Dahlin DC. Grading of bone tumors. Semin Diagn Pathol. 1984 Aug;1(3):165-72. PMID: 6400634.