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Adult Langerhans Cell Histiocytosis a Rare Entity in Cervical Spine Compression: Case Report

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Abstract:-

- ***Case***

A 43 year old male presented with complaints of neck pain radiating to bilateral upper limb with weakness of bilateral upper limbs. Examination revealed a loss of cervical lordosis with restricted range of motion of the cervical spine .Neurological examination revealed a weakness of finger abduction and adduction. Blood investigations showed an elevated erythrocyte sedimentation rate, while the rest were within normal limits.MRI scan of the cervical spine showed an hyperintense lesion of the first thoracic vertebral body. CT guided biopsy was attempted followed by open biopsy of the lesion and specimen was sent for histopathological examination which was later reported as Langerhans cell histiocytosis and the patient was treated with chemotherapy for the same.

- ***Conclusion***

Despite the rarity of the condition, the possibility of LCH should be taken into account in these circumstances. This case shows the role of chemotherapy in management and draws attention towards early detection in order to prevent neurological complications with respect to the rarity of the disease and the site of the bone involvement.

Keywords:- *Langerhans Cell Histiocytosis, Chemotherapy.*

I. INTRODUCTION

Langerhans cell histiocytosis is a rare, non-neoplastic disease of unknown etiology, usually found in patients younger than 20 years. It is characterized by a clonal proliferation of specialized dendritic cells. LCH may be accompanied by the potential risk of associated complications, depending on location and spread of the lesions. The accumulation of Langerhans-cells in a bone causes the classic lytic bone lesions, which may lead to neurological deficits .Although it frequently manifests in children as cranial lesions, it can also manifest as a spinal lesion, with the thoracic spine being the most typical location.[1] Here, we describe a case of cervical langerhans cell histiocytosis who presented with neurological deficits. The patient was managed with chemotherapy and is free of recurrence after 1 year.

II. CASE PRESENTATION

A 43 year old male presented with complaints of neck pain radiating to bilateral upper limb with weakness of bilateral upper limbs since 1 year . On examining the patient, loss of cervical lordosis was noted with no palpable swelling or deformity with restricted range of motion of the cervical spine .Neurological examination revealed a weakness of finger abduction and adduction . Patient presented with an MRI scan of the cervical spine which showed a hyperintense lesion of the first thoracic vertebral body with altered signal intensity in the anterior epidural space and the paraspinal soft tissues.

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Fig 1: MRI cervical spine sagittal sections which show a hyperintense lesion of the first thoracic vertebrae in T2 weighted images.

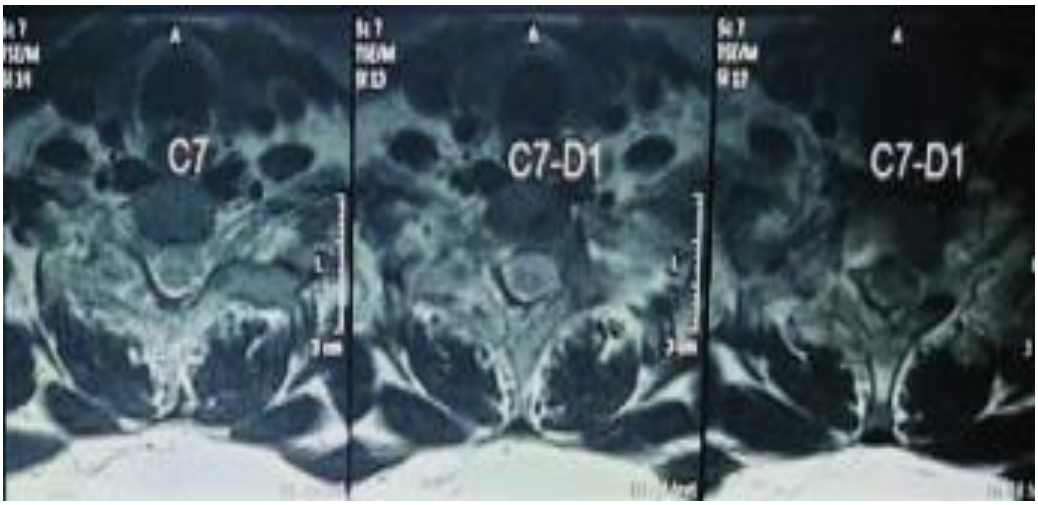

Fig 2: MRI cervical spine axial sections which show altered signal intensity in the anterior epidural space and the paraspinal soft tissues.

Based on the above given findings based on the imaging and co relating with the examination findings , the differential diagnosis which were thought of were tubercular lesion of the cervical spine, secondaries involving the cervical spine and primary lymphoma of the cervical spine.

Keeping the above mentioned differential diagnosis in hindsight , the patient was evaluated for the above. Chest x ray was done which showed no radiological abnormalities.

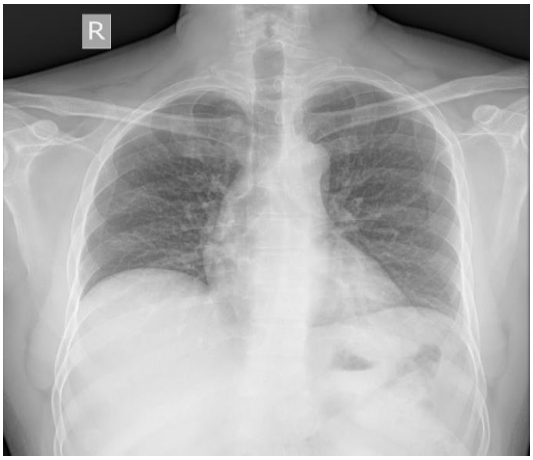

Fig 3: Chest x-ray of the patient , which showed no radiological abnormalities.

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PET CT scan was done to look for secondaries which showed increased uptake of FDG at the lytic lesion at C7 and D1 vertebral levels along with epidural and paravertebral soft tissues.

CT guided biopsy was planned for the lesion in order to obtain a sample for histopathology from the involved vertebral level which turned out to be inconclusive. The CT scan of the cervical spine showed lytic lesions of the C7 and D1 vertebrae.

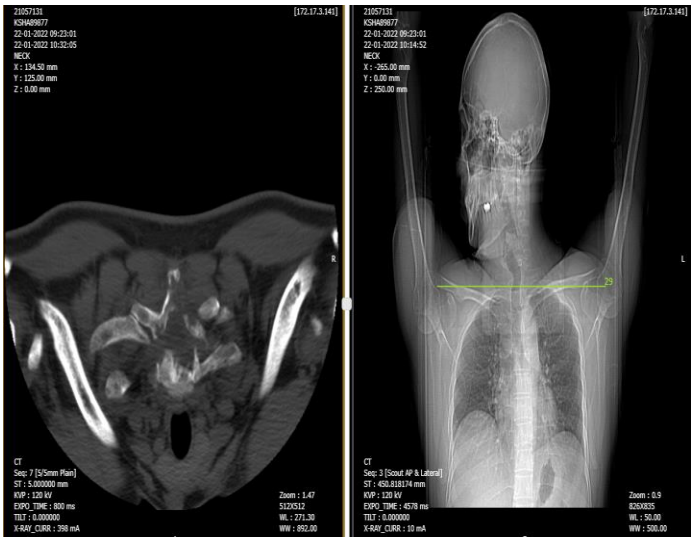

Fig 4: CT scan of the cervical spine showing lytic lesion at C7 and D1 level.

An open biopsy was performed at C7 level and the vertebral body bony elements were sent for histopathological examination.

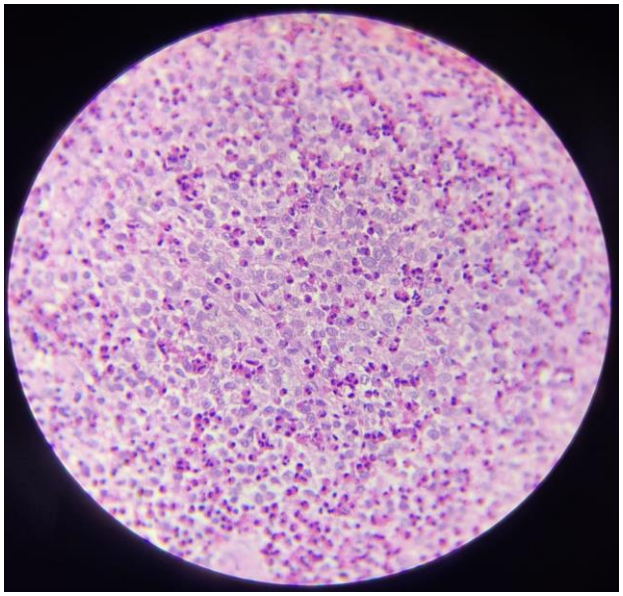

Fig 5: Microscopy image of the biopsy specimen showing epithelioid histiocytes

The histopathology specimen showed irregular bony trabeculae with infiltrative lesions consisting of intermediate sizes of histiocytes in aggregates having longitudinal grooving along with plenty of eosinophils forming microabscess.

Immunohistochemistry staining was positive for CD1a and S-100 which furthermore confirmed the diagnosis of LCH.

Patient was planned for and started on a chemotherapy regimen with Vinblastine along with Prednisolone for a period of 6 weeks.

He was re-evaluated after 3 months after initiation of chemotherapy and was found to be symptomatically better with no further progression of neurological deficits.

He is currently 1 year post chemotherapy and is symptomatically better with no further deficits and is able to carry out his routine activities without any restrictions.

III. DISCUSSION

Histiocytic disorders are characterized by an aberrant buildup of mononuclear phagocytic system cells in children and adults. LCH can involve a single organ (single-system LCH), which can be a single site (unifocal) or several sites (multifocal); or multiple organs (multisystem LCH), which can involve a limited number of organs or be diffused. The involvement of certain organs such as the liver, spleen, and hematological system divides multisystem LCH into high-risk and low-risk categories, where risk implies the chance of disease-related death.[2]

One of the most commonly impacted systems is the bone . Although some bone lesions are asymptomatic, others cause pain and a soft elevated mass in a specific location of the bone . A single "punched out" lytic lesion of the skull, symmetrical fattening of the anterior and middle vertebral column ("vertebra plana"), and endosteal scalloping of the long bones are typical radiographic findings.

The skull is the most commonly involved region, followed by the spine, extremities, pelvis, and ribs.

Therapeutic options for solitary lesions of the spine are dependent on location, symptoms and the presence or absence of neurological malfunction and range from clinical/radiological monitoring to cervical or lumbar orthoses.

CT-navigated intralesional infiltration of corticosteroids, surgical curettage and bone grafting, or even radical surgical options such as the resection of the affected bone.

Conservative treatment options, such as observation, bracing or immobilization seem to have a positive impact on the outcome especially in paediatric patients without neurologic deficit.

Green et al. [3] successfully managed two patients with neural deficit using radiation therapy. They suggested that surgical decompression of these lesions was probably not necessary because any neural deficit would quickly disappear after biopsy and irradiation. In spite of the facts, there is still no clear evidence as to whether radiation therapy is beneficial to the patients.

Despite the fact that chemotherapy has been proposed as a systemic treatment for multi-site bone lesions and multiple system involvements, there is no evidence that chemotherapy is a good option in the management of LCH of the spine in children with soft tissue extension, particularly those with neurological deficits. In this work, we demonstrated that chemotherapy was capable of rapidly reducing intraspinal soft tissue mass and relieving local and radicular pain, even in the presence of spinal cord and nerve root compression.

Tanaka et al. [4] employed prednisone and methotrexate to treat two cases of atlas LCH, and they appear to be both safe and efficacious. Tan et al.[5] used only prednisone to treat four children with cervical LCH; all patients healed completely and retained normal cervical spine function. Womer et al. [6] discussed treating low-risk LCH with alternate-day prednisone and weekly methotrexate. These researchers came to the conclusion that low-dose chemotherapy was both safe and effective. In our case we found chemotherapy to be an effective modality in treating this lesion.

IV. CONCLUSION

LCH continues to provide diagnostic and therapeutic challenges. Although it is uncommon in the cervical spine, it is important to keep this disease in mind. Imaging should be thoroughly examined before excluding other explanations. Conservative and operative interventions should be decided upon keeping in mind the extent of the lesion and the neurologic deficits.

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CONSENT

The patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.