Langerhans Cell Histocytosis in Cervical Spine: A Rare Case Report

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Abstract

Langerhans cell histocytosis (LCH) in spine is a benign disorder that rare in adults and mainly affects children. We report the case of a 16 year-male presented to us with severe pain in nape region and upper limb radiculopathy along with upper limbs weakness for 4 months. He had history of fall injury 6month ago. Radiological investigation revealed to have a solitary osteolytic lesion with pathological fracture at C6 vertebral body. The patient underwent surgical management in the form of excision of lesion, anterior C6 Corpectomy with C5-C6, C6-C7 discetomy and reconstruction with titanium cage and plating. Patient had immediate relief of nape region pain and radiculopathy. The accurate diagnosis of LCH was founded on histological examination.

Key words: Langerhans cell histocytosis, cervical spine, anterior cervical corpectomy with discetomy and fixation

Introduction

Langerhans cell histocytosis (LCH) is a rare disease. It results from abnormal proliferation of Langerhans cell, which results in the formation lesion primarily in the skin, bone, liver, spleen and lymph nodes. [1, 2] LCH was named by Paul Langerhans's name who first discovered the epidermal dendritic cell in 1868. [3] There are some studies have described genetic mutation of B-RAF gene [4] and BRAF V600E gene [5] are the causes of LCH. Even though the exact causes of LCH are still unknown. Almost 80% of patients presents before the age of 10 year, making this is a childhood diseases. [6] LCH is more common in children, rare in adult, and ratio of male to female is 1.2-2:1.[7, 8] Spine involvement has been found in approximately 7-15% of cases (9) with a predilection for the thoracic spine (54%) followed by the lumber (35%) and the cervical (11%).[10] There are no precise guidelines for diagnosis and treatment of an adult despite the clinical features and management of the diseases are well known in children.[11]

We report a case of LCH at C6 vertebrae an adult patient who presented to us with nape region pain along with pathological fracture and upper limbs radiculopathy treated with anterior excision of lesion, anterior corpectomy with discetomy and reconstruction done with titanium cage and plating.

Case report

A 16 year male admitted to neurosurgery department with complain of restricted neck mobility, nape region pain radiating to B/L shoulder and upper limbs, weakness and numbness of upper both limbs for the past 4 months. He had history of fall injury 6 month ago following injury his pain had significantly increased and there was no significant history of fever, weight loss, and night sweats and decreases appetite and pain at other site of body. A physical examination demonstrated localized tenderness over C6 spinous process, restricted neck mobility, numbness and weakness of both upper limbs. Laboratory tests, including full blood count, serum electrolytes, and renal function and liver function tests did not reveal any abnormalities. Erythrocyte sedimentation rate and C-reactive protein were increased to varying degrees. Magnetic resonance imaging (MRI) revealed C6 osteolytic destruction with increased STIR signal in the body adjacent posterior elements, posterior convex vertebral margin associated with homogeneously enhancing anterior epidural soft tissue obliterating anterior thecal sac causing central canal stenosis and larger similar pre-paravertebal

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soft tissue at C5,C6,C7 vertebral level (Figure 1). Anterior cervical C6 corpectomy and discectomy was done after lesion excising for biopsy and reconstruction was done with titanium cage and plating (Figure 2).

Histological examination of the biopsy sample showed proliferation of Langerhans cells in sheets and associated with others cell such as eosinophils, lymphocytes, macrophages, and plasma cell and it was reported to be Langerhans cell histocytosis (Figure 3). Immediately post surgery, his pain and numbness of upper limbs decreased significantly. He was mobilized with cervical collar on the next day after surgery and discharged after 5th day of surgery. He had no any neurological symptoms during follow up period.

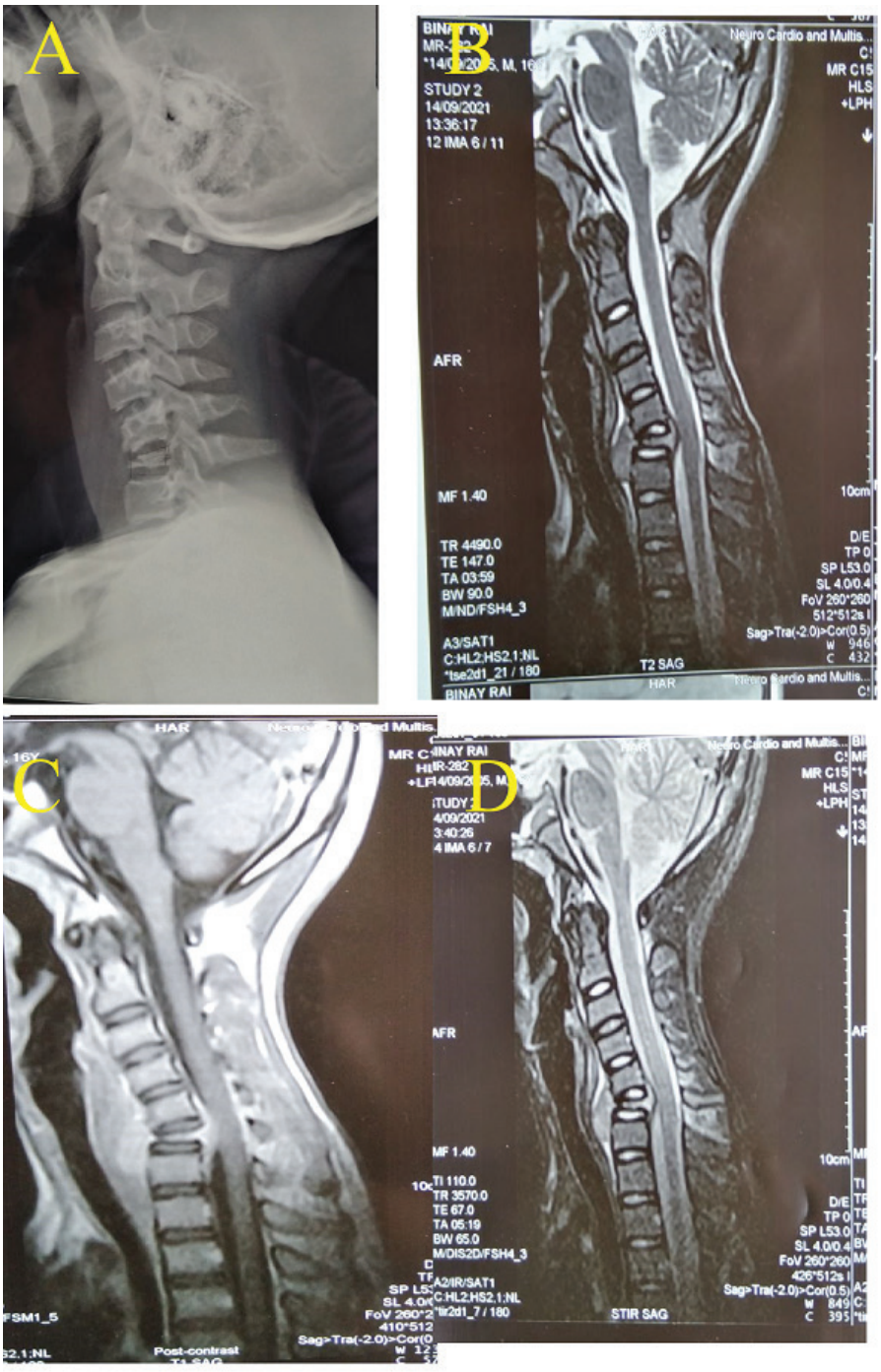

Figure 1: C-spine x-ray view showing osteolytic destruction of C6 vertebra (A) and Magnetic resonance imaging (MRI) revealed C6 osteolytic destruction with hyper intense lesion in T2 sagittal, T1 sagittal and STIR sagittal images in C6 vertebral body (B, C, D)

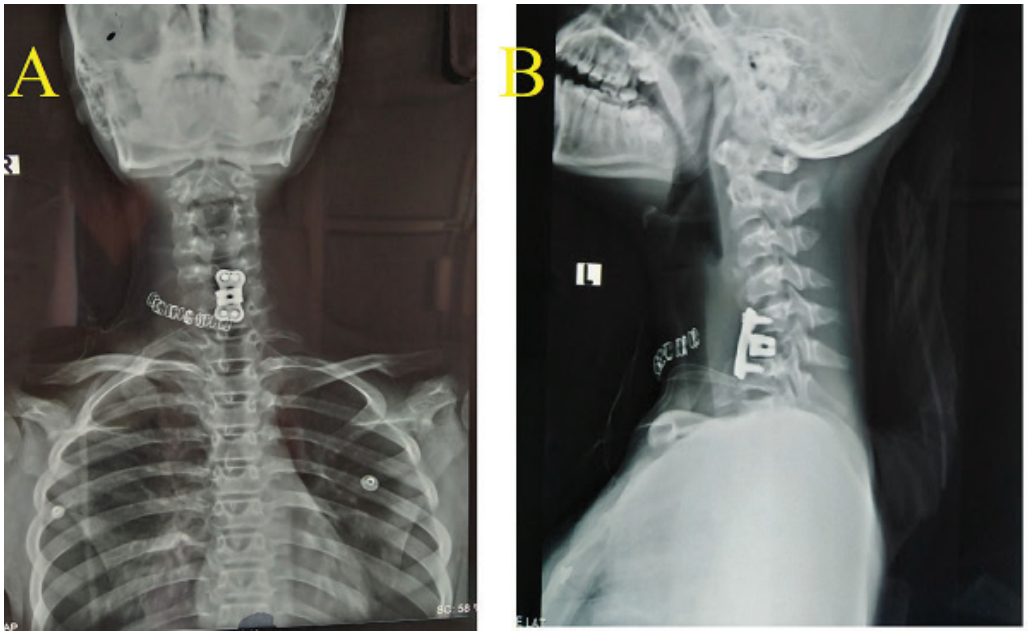

Figure 2: Anterior cervical C6 corpectomy and discectomy was done after lesion excising for biopsy and reconstruction was done with titanium cage and plating

Low magnification (H and E x 100) overviews of discrete nodules formed by cells separated by fibrous tissue (A), High Magnification (H and E x 400) shows cells with eosinophilic cytoplasm (B), High Magnification (H and E x 400) shows lymphocytes, eosinophils and plasma cell in between (C) and High Magnification (H and E x 4) shows lymphocyte liked cells (D).

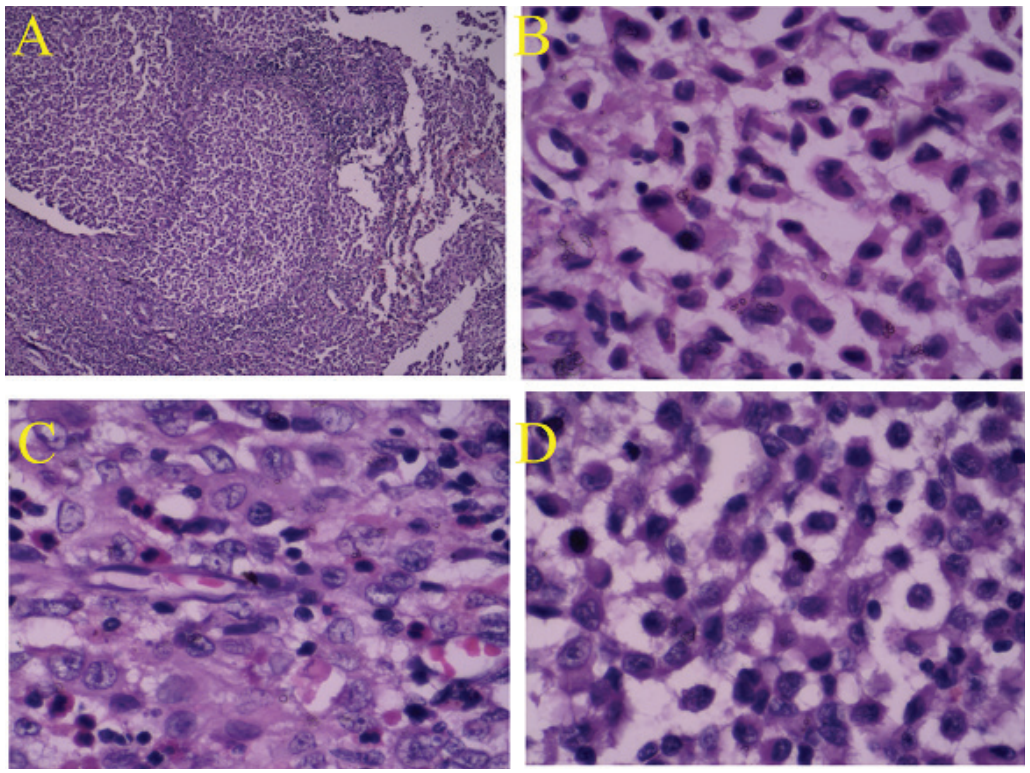

Figure 3: Low magnification (H and E x 100) overviews of discrete nodules formed by cells separated by fibrous tissue (A), High Magnification (H and E x 400) shows cells with eosinophilic cytoplasm (B), High Magnification (H and E x 400) shows lymphocytes, eosinophils and plasma cell in between (C) and High Magnification (H and E x 4) shows lymphocyte liked cells (D)

Discussion

Langerhans cell histocytosis is a disease of unrecognized physiological mechanism as well as rare disease, and identified by a clonal proliferation of Langerhans histocytes cell as well as eosinophils, macrophages, lymphocytes, and occasionally multinucleated giant cell.[12, 13] Langerhans cell histocytosis mainly affects children with a peak incidence of 1 to 5 year and incidence of Langerhans cell histocytosis is 1-2 cases per million per year.[14] In our study, age of patient was 16 year.

The clinical symptoms of LCH differ such as skin rash, fever, anemia, thrombocytopenia, hepatospenomegaly, lymphadenopathy, and which can lead to multi organ failure and often death within 1-2 years.[15] LCH can affect almost every organ of the body.[16] The skeletal lesion of LCH are most common in the skull (26%). and less common in the vertebrae (7%), ribs (12%), upper and lower jaws (9%), and bone of extremities (11%). [11,17] LCH mainly affects the vertebral bodies in the spine, with most common in the thoracic spine (54%), followed by the lumbar (35%) and cervical spine (11%).[18] Patients with cervical spine or any vertebral body involvement usually have local pain, limited range of motion, or motor or sensory dysfunction.[19] In our patient C6 vertebral body was involved without involvement of posterior structures and radiological investigation including magnetic resonance imaging (MRI) revealed C6 osteolytic destruction reported hyper intense lesion in T2 sagittal images and hyperintense in T1 sagittal images in C6 vertebral body, which is similar to other studies.[20]

The various treatment options including observation, non-steroidal anti-inflammatory drugs, chemotherapy, radiotherapy, intralesional steroid and surgery in the form of excision of the lesion and reconstruction of spine have been described.[11] The definite guidelines or indication of radical surgery have not reported well. We decided to perform radical surgery because patient had radiculopathy, motor and sensory deficits. Therefore, surgical treatment in the form of anterior cervical C6 corpectomy and discectomy was done after lesion excising for biopsy and reconstruction was done with titanium cage and plating. This is a single case study, therefore further study with large number of cases should be done to establish proper guidelines and indication for surgical treatment of LCH of adult cervical spine. A definitive diagnosis of LCH was made by histopathological study of the biopsy of lesion.

Conclusions

Surgical treatment in the form anterior cervical corpectomy with discectomy and titanium cage plus plating for LCH in adults should be considered in patients with refractory nape pain and with pathological fracture, neurological deficits

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