LANGERHANS CELL HISTIOCYTOSIS OF THE SCAPULA - DIAGNOSIS & TREATMENT OPTIONS

HISTIOCITOSE DE CÉLULAS DE LANGERHANS DA ESCÁPULA - DIAGNÓSTICO E OPÇÕES DE TRATAMENTO

HISTIOCITOSIS DE CÉLULAS DE LANGERHANS DE LA ESCÁPULA - DIAGNÓSTICO Y OPCIONES DE TRATAMIENTO

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ABSTRACT

Langerhans cell histiocytosis (LCH) is characterised by an abnormal histiocytic accumulation in tissues such as the lung, spleen, bone marrow, skin, central nervous system, liver and lymph nodes, causing focal or systemic effects. No specific clinical & radiographic presentation of LCH is described in literature. This poses a diagnostic dilemma for surgeons. The scapula is the site of 3% of bone tumours, while for LCH it is the least common site. In a 10-year-old boy with isolated lesion of the scapula with no other systemic involvement, and no specific finding in MRI or CT scan of scapula, diagnosis was confirmed on biopsy. Division into single and multi-system disease is paramount in treatment, given that it is a single system disease. The patient improved clinically on follow-up of 2 years. The scapula is one of the rarest site of LCH, and because various lesions mimic each other, a biopsy is always required, with immunohistochemistry for CD68 & S-100. This was only a single system disease, so conservative management was performed, and the patent improved clinically.

Keywords: Histiocytosis, Langerhans-cell; Scapula; Immunohistochemistry.

RESUMO

A histiocitose de células de Langerhans (HCL) caracteriza-se por acúmulo anormal de histiócitos em tecidos como pulmão, baço, medula óssea, pele, sistema nervoso central, fígado e linfonodos, causando efeitos focais ou sistêmicos. Nenhuma apresentação clínica e radiográfica específica da HCL está descrita na literatura. Isso impõe um dilema diagnóstico para os cirurgiões. A escápula é o local de 3% dos tumores ósseos, ao passo que é o lugar menos comum para a HCL. Em um menino de 10 anos de idade, com lesão isolada na escápula e sem outro envolvimento sistêmico, sem achados específicos na RM ou na TC da escápula, o diagnóstico foi confirmado pela biópsia. A divisão entre doença isolada e de múltiplos sistemas é fundamental para o tratamento, considerando-se que este caso é uma doença de um só sistema. O paciente teve melhora clínica no acompanhamento de dois anos. A escápula é um dos locais mais raros de ocorrência da HCL, e como as lesões mimetizam umas às outras, sempre é preciso realizar biópsia por imuno-histoquímica para CD68 e S-100. Esta doença atingiu apenas um sistema, levando ao tratamento conservador e o paciente apresentou melhora clínica.

Descritores: Histiocitose de células de Langerhans; Escápula; Imuno-histoquímica.

RESUMEN

La histiocitosis de células de Langerhans (HCL) se caracteriza por la acumulación anormal de histiocitos en tejidos como pulmón, bazo, médula ósea, piel, sistema nervioso central, hígado y linfonodos, causando efectos focales o sistémicos. Ninguna presentación clínica y radiográfica específica de la HCL está descrita en la literatura. Eso impone un dilema diagnóstico para los cirujanos. La escápula es el local de 3% de los tumores óseos, al paso que es el lugar menos común para la HCL. En un niño de 10 años de edad, con lesión aislada en la escápula y sin otro compromiso sistémico, sin hallazgos específicos en la RM o en la TC de la escápula, el diagnóstico fue confirmado por la biopsia. La división entre enfermedad aislada y de múltiples sistemas es fundamental para el tratamiento, considerándose que este caso es una enfermedad de un único sistema. El paciente tuvo mejora clínica en el acompañamiento de dos años. La escápula es uno de los locales más raros de ocurrencia de la HCL, y como las lesiones mimetizan unas a otras, siempre es preciso realizar biopsia por inmunohistoquímica para CD68 y S-100. Esta enfermedad alcanzó a sólo un sistema, llevando al tratamiento conservador y el paciente presentó mejora clínica.

Descriptores: Histiocitosis de células de Langerhans; Escápula; Inmunohistoquímica.

INTRODUCTION

Langerhans cell histiocytosis (LCH) is characterized by an abnormal histiocytic accumulation in tissues such as the lung, spleen, bone marrow, skin, central nervous system, liver and lymph nodes, causing focal or systemic effects.[1] It is a rare disease, and its aetiology is still under observation. Newer studies have suggested abnormal immune regulation, and some viruses, like the Epstein Barr virus, as an etiological agent. Eosinophilic granuloma (EG) is a variant that commonly presents with skeletal manifestations. No specific clinical & radiographic presentation of LCH is described in the literature, and this poses a diagnostic dilemma for surgeons.[2-5]

Study conducted at the University College of Medical Sciences, Department of Orthopaedics, and Guru Teg Bahadur Hospital, Dilshad Garden, Delhi, India.
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The bones are commonly affected, especially the flat bones, the spine and the long bones. Lesions such as "vertebra plana" or solitary lytic lesion of the skull may suggest LCH in children, while other lesions may be confused with malignant tumor or osteomyelitis. The scapula is the site of 3% of bone tumors. Benign tumors like osteochondroma, osteoid osteoma, fibromatosis, osteoblastoma, and aneurysmal bone cyst are more common. Malignant tumors such as chondrosarcoma, Ewing's sarcoma, osteosarcoma, and lymphoma may also mimic LCH. The scapula is one of the rarest site of LCH. Here, we present a rare case of isolated LCH scapula with no other lesion in the body, and with a benign course.

CASE REPORT

A 10-year-old boy was reported in our institute in 2014, with chief complaints of pain in the right shoulder and painful restriction of movement of the right upper limb since one year previously. Patient was apparently in good health until 2013, when he suffered a minor fall while playing, upon which he was taken to an orthopedic doctor in his home town, where he was diagnosed as having no bony injury. The patient was given an arm sling, and was asymptomatic immediately afterwards. In 2014, the patient again complained of pain in the right shoulder with insidious onset, gradually progressive, aggravated on activity with the upper limb, and partially relieved with analgesics, with no swelling. The patient received analgesics on and off for one year. The pain further increased in the last 6 months, and the patient was referred to GTB hospital, where a biopsy was performed. On general physical examination he was conscious, oriented, vital stable, and afebrile with no lymphadenopathy. On local examination, there was a diffuse swelling over the right scapula in the posterior lateral view; the overlying skin was mobile, there were no dilated veins, abduction on the right side was limited, and the remaining range of motion was comparable. (Figure 1) On palpation there was an area of swelling over the scapula of approximately 4x4 cm, with poorly-defined margins, and with varying consistency, and thickening of the lateral border of scapula. Hematological investigation showed: Hemoglobin - 10.9 gm%, TLC- 7,900/mm^3, DLC- 75(N)/25(L), ESR - 02 mm/hr. X-rays of the right shoulder were performed in the AP & lateral views, showing, at first presentation in 2013, a lytic geographical lesion in the scapular neck region, which progressed over the years into a moth-eaten type picture involving the entire scapula. (Figures 2 and 3) MRI & CT scans of the right shoulder showed infiltration of the scapula with some soft tissue edema on MRI. (Figure 4).

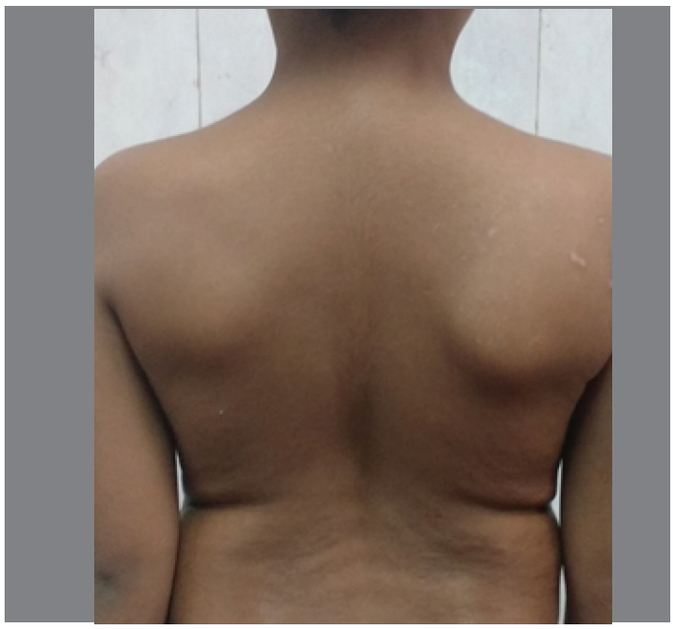

Figure 1. Clinical photograph showing swelling in posterior aspect of scapula.

The remaining x-ray investigations: Chest – PA view, skeletal survey, Bone scan (Figure 5) & USG Abdomen showed no other lesions in the body. Biopsy was taken, which revealed fibrocartilaginous tissue with numerous histiocytes with prominent grooves, and mild to moderate pleomorphic with eosinophils. On Immunohistochemistry, CD68 and S-100 were positive. These findings were suggestive of a means distinguishing features of LCH.

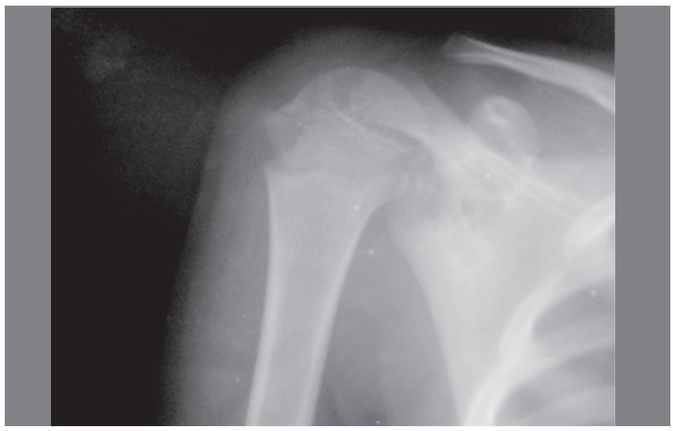

Figure 2. X-ray AP view scapula showing geographical lesion scapular neck.

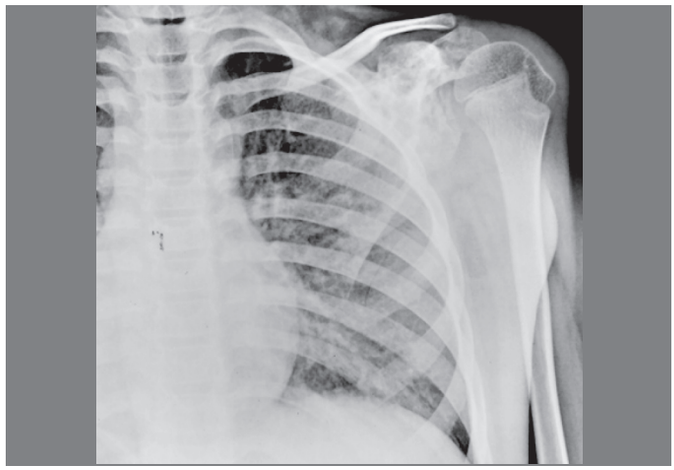

Figure 3. X-ray showing complete involvement of scapula neck and acromion..

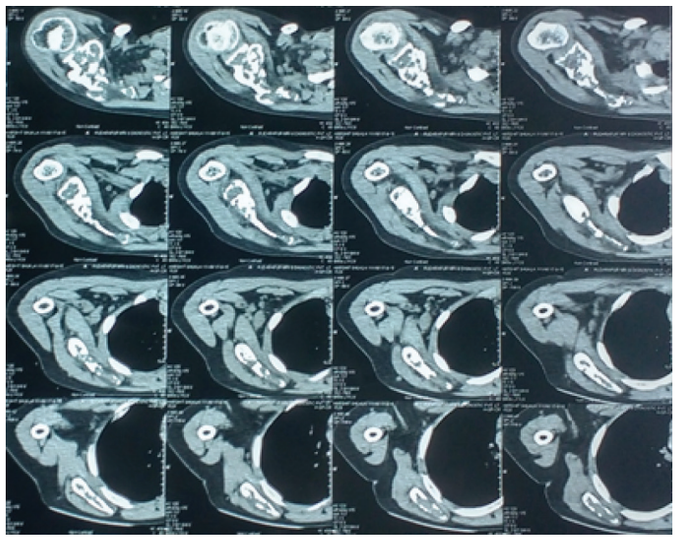

Figure 4. MRI showing infiltration of scapula with soft tissue edema of the right shoulder.

On further investigation, we were not able to find any other injury to the skeletal system or systemic circulation. The patient was treated as having a single system disease and after biopsy, the patient was closely observed. As of the last follow-up at 2 years, the patient is improving clinically, the pain has reduced significantly and no other focus of disease is observed in any other part of the body. (Figure 6)

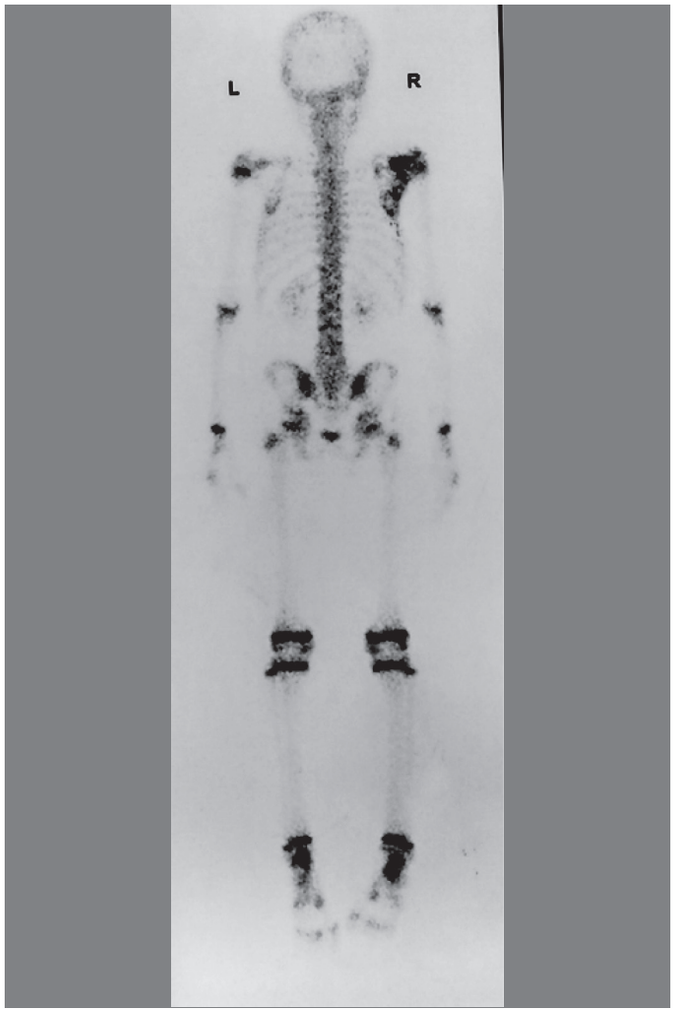

Figure 5. Bone scan shows no other active lesion in the body.

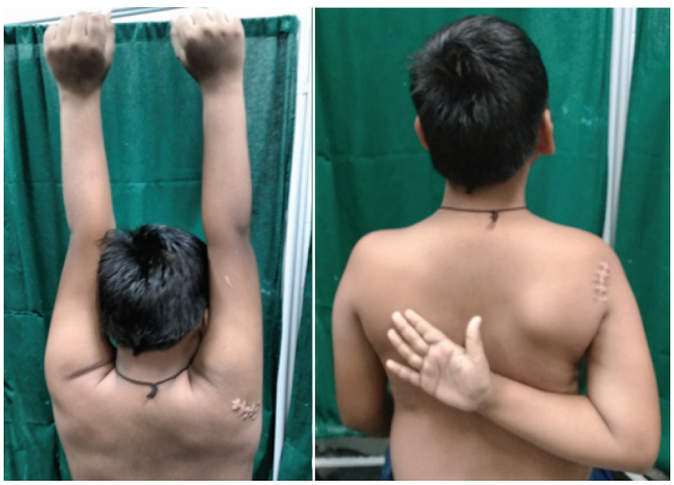

Figure 6. Clinical photograph showing complete range of motion at 2-year follow-up.

DISCUSSION

Langerhans cell histiocytosis (LCH) is a versatile mimicker that is difficult to diagnosis.[6] The disease course has a spectrum from spontaneous resolution to progressive multisystem disorder, and may also involve potential life-threatening complications.[6-9]

The most common clinical presentation of LCH in children is pain. They mostly develop localized soft tissue swelling, often with bone defects that are sharply demarcated on palpation. The diagnosis is made through biopsy of the lesion.

Pathological specimen shows bone infiltration by characteristic histiocytic cluster with an admixture of morphologically related giant cells, eosinophils and lymphocytes. Electron microscopy demonstrates specific Langerhans granules (Birbeck granules) in the histiocytes. Immunohistochemistry shows strong S-100 protein, HLA-DR and CD-1a surface antigen positivity by the histiocytosis X cells.[10,11] It is believed by many authors that a definitive diagnosis can be established by routine haematoxylin and eosin morphology especially when correlated with radiological and clinical data.[12]

The treatment of Langerhans cell histiocytosis (LCH) is still controversial and has changed over the past decades. Initially, it was thought to be granulomatous, inflammatory, or infectious disease and was treated with anti-inflammatory agents, steroids and antibiotics. With advances in pathogenesis, the trend now is towards radiation therapy and cytotoxic chemotherapy. While varying degrees of success have been reported, it should be evident that only once the aetiology and pathogenesis have been resolved can a definitive therapy be envisioned.

Currently, treatment of LCH is divided into single system and multiple system disease. Single system disease (usually bone, lymph node or skin) generally involves benign course, with high spontaneous remission and favorable outcome over a period of time in most cases.[13]

Bony lesions usually do not require treatment other than biopsy to confirm diagnosis, or curettage at the time of initial biopsy. Further local therapy is recommended only if weight-bearing bones (with a risk of spontaneous fracture) are involved, or if the lesions cause pain or could result in unacceptable dysfunction or deformity. Intralesional infiltration of steroids (40-200 mg methylprednisolone) was recently reported as an effective and safe treatment.[14] Radiation therapy as a treatment is condemned nowadays, as the LCH single system is found to be self-limiting. Also, there have been reports of a 5% risk of malignancy after irradiation.[15] In circumstances when the consequences of the disease threaten the function of a critical organ (e.g. lesions surrounding the optic nerve or spinal cord) and local infiltration with corticosteroids is not feasible, treatment with low dose radiotherapy (6-1O Gy) is warranted.

In multiple system disease when multiple bony lesion are present or risk organs are involved such as the bone marrow, lymph node, spleen, or liver. Systemic chemotherapy is warranted. Newer treatments with gene therapy and bone marrow transplantation are also considered these days.

It is very important to look at the patient's profile and the general course of disease to decide on the management plan for the patient, as there is no specific protocol. Also, the scapula is one of the rarest sites of involvement of LCH, so it is important to keep an open mind, while differentiating between scapular tumors.

CONCLUSION

Currently, in reactive disease like LCH, the response is defined as a measurable resolution of symptoms and signs and the prevention of permanent consequences of the disease. As in this rare case only single system is involved, we have adopted a conservative approach. At the two-year follow-up, despite the stagnant radiological appearance, there is resolution of the clinical signs and symptoms clinically, and the child is able to do carry out his normal daily activities.

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