Title: Case Report of Langerhans Cell Histiocytosis (LCH) in the Scapula Presented to Bouali Sina Hospital, Sari, Iran at January 2020

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Abstract

Langerhans cell histiocytosis (LCH) is an uncommon hematological disorder-affecting infant and young child.LCH is a rare disorder of the reticuloendothelial system associated with proliferation of Langerhans cells and mature eosinophils. LCH can involve any bone, but the most common are pelvis, ribs, skull, long bones, vertebra, and facial bones. In this article, we report a case of LCH in a 15-months-old child involving the scapula and round shape swelling on left scapula with no erythema or tenderness and normal passive range of motion of shoulder and also aggressive periosteal reaction led to the diagnosis of LCH. This patient undergone surgery and received chemotherapy with no complications or any recurrence after two years follow up

The purpose of reporting this case is to discuss clinical and radiological and histopathology features of LCH and role of doctors in diagnosing and managing such lesions.

Keywords: Langerhans cell histiocytosis, LCH, Scapula

Introduction:

Langerhans cells are macrophages body organs whom proliferation is in an abnormal way, and they thought to be responsible for a disease called Langerhans Cells Histiocytosis (LCH) which can damage bone, skin, lymph nodes, lung, liver, spleen and bone marrow. (1,2)

LCH happens in any age group but it has a peak in 5-15 years. (3,4)

In flat bones like skull there is a variant of LCH called Eosinophilic granuloma (EG) which usually presents with skeletal features. (5)

Diagnosis of LCH is based on biopsy and histological study. Although children usually present with pain and it is the most common symptom, but it has no specific clinical features and radiological appearance. (6)

Outcome of LCH varies from spontaneous resolution to progressive multisystem disorder. (7)

In this case we are presenting a 15 months old boy with LCH of scapula

Case Repot:

A 15-month old baby boy was presented to pediatrics clinic with a mass on the posterior aspect of his left shoulder which his mother had found 1 month earlier. He had a history of trauma to the shoulder from falling down the chair 2 months preferably.

They didn't detect any symptoms of fever, chill, night sweats or weight loss and his mother didn't attack any abnormality in using the left arm.

He had no other past medical history and his family were healthy without any history of malignancy.

On physical examination there was a round shape swelling on left scapula with no erythema or tenderness, passive range of motion in both shoulders were similar.

Results of paraclinical examinations are shown in table 1.

Table 1- results of paraclinical studies

Hemoglobin	11.3	CRP	23
WBC	4570	ESR	7
Neutrophil	32.6%	Creatinine	0.42
Lymphocyte	55%	Calcium	9.2
Platelet	378000	Phosphor	4.6
AST	33	Magnesium	1.75
ALT	8	Serum Albumin	3.76
Alkaline phosphatase	777	Total protein	6.3
LDH	328	Bilirubin Total(Direct)	0.1 (0.04)

On radiography, there was a round lucent structure in left scapula (figure-1).

Computed Tomography (CT scan) showed a soft tissue mass in body of left scapula with expansion to anterior cortex and disruption of posterior cortex, this lesion expanded to bulk of Infraspinatus and teres minor muscles (Figure-2).

Magnetic Resonance Imaging (MRI) with and without Gadolinium injection was performed that revealed a heterogenous enhancing expansile mass (33*26*22 mm) in the left scapula with marked peripheral muscles enhancement and edema and a few lymph nodes in the left axillary region without intrathoracic invasion or involvement of humorous(Figure-3).

Histopathology reveal a mesenchymal neoplasm composed of diffuse infiltration of histiocytes with lobulated, indented nuclei and eosinophilic cytoplasm admixed by many eosinophils (Figure-4).

Abdominopelvic sonography was normal.

Bone marrow aspiration showed normal cellular marrow and in biopsy of bone marrow, there were Langerhans cells infiltration.

He undergone surgery to remove the tumor and a little of healthy tissue around it.

The patient received 6 weeks of vinblastine 0.2 mg/kg body weight along with oral prednisolone 1 mg/kg body weight.

A repeat examination and imaging were done 3 months after completion of chemotherapy did not reveal any activity noted previously.

The patient is currently being followed up for last 1 years without any recurrence.

Discussion

LCH is a disease that happens in children more than other age groups, male gender is more affected, and it occurs most in age 1-3 years. (8-9) The annual incidence of LCH has been estimated to be 2 to 10 cases per 1 million children aged 15 years or younger (10)

Head and neck are the most common area that this disease presented in. (11)

We can use imaging and they play a key role to achieve diagnosis and in management of LCH, especially in isolated bone lesions. Computed tomography (CT) also can be used as the initial modality of diagnosis. Although LCH has no pathognomonic radiologic appearance, it has characteristics such as well-defined lytic lesion with a punched-out appearance .(12)

Before confirming of LCH as a diagnosis for a isolated bone lesion, a complete evaluation of skeletal system should be done, searching to find other bone lesions. (13-14)

We should try to confirm diagnosis of LCH by biopsy if possible, to rule out other bony lesions with a similar radiologic appearance. This disease can be self-limited and regress spontaneously, or it can be progressive, so the course is unpredictable. Therefore, for choosing treatment options routine imaging must be done and patient must be followed up long time after resolution.(12)

Because of its low prevalence and variable location of bone lesions, and because of variability in severity and different types of LCH, there is no a definite guideline for treatment of this disease.

Chemotherapy and other aggressive treatment options can be used in multisystem form of LCH. Bezdjian et al. reviewed 201 patients from 45 published studies of isolated LCH bony lesions and formulated a systematic algorithm for diagnosis, investigations and management (15)

Age of diagnosis of isolated LCH varies from 2 weeks to 17 years with a mean age of 8.1±4.3 years. The usual symptoms were swelling (64%), pain and swelling (18%), and pain only (9%) while some patients even presented with torticollis, paresthesia, and bleeding. LCH bony lesions were located in the skull (61%), orbit (24%), cervical spine (8%), and mandible (4%) and each of these sites may be a part of either unifocal unisystem, multifocal unisystem, or multisystem disease, but isolated solitary lesions of the orbit was not common (15).

Our patient was a 15-month old baby and the disease was presented only with swelling at the posterior aspect of his left shoulder area without evidence of any other foci of eosinophilic granulomas anywhere else in the body.

In the literature there are different kinds of management of solitary bone lesion and many authors suggest treatment with different modalities, such as observation, surgical curettage, radiation therapy, steroid injections, and chemotherapy, or a combination of two or more, with good prognosis (16-17).

Because of the similarity of this disease to neoplasia, the hypothesis highlighting led to establish trials using chemotherapy and radiation therapy as one of the ways to eradicate the lesion.

Radiation and chemotherapy have been used for unifocal lesions that fail spontaneous resolution and are difficult to resect surgically (18-19).

Furthermore, the rate of recurrence of isolated bone lesion is very low despite modality of treatment (20).

Intra lesion injection of steroids leads to a high rate of recurrence, so this treatment option has been avoided (10-15).

Radiation, even though administered to the localized area of involvement, has its accompanying complications, such as: skin necrosis, hair loss, neurological damage (21)

By considering benefits and side effects of each treatment, we choose surgery followed by chemotherapy for our patient. Now he is a healthy boy without any sequelae of disease and he has no evidence of recurrence.

Conclusion

Diagnosed as an isolated solitary bone lesion of LCH in scapula, our patient after mass removal received vinblastine chemotherapy without evidence of any complications and without any recurrence over the 2 years of follow up.

In our opinion, chemotherapy is a relatively safe and

effective treatment option after surgery in pediatric patients and may be considered adjuvant modality of choice in isolated solitary as well multisystem LCH.

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Figure-1

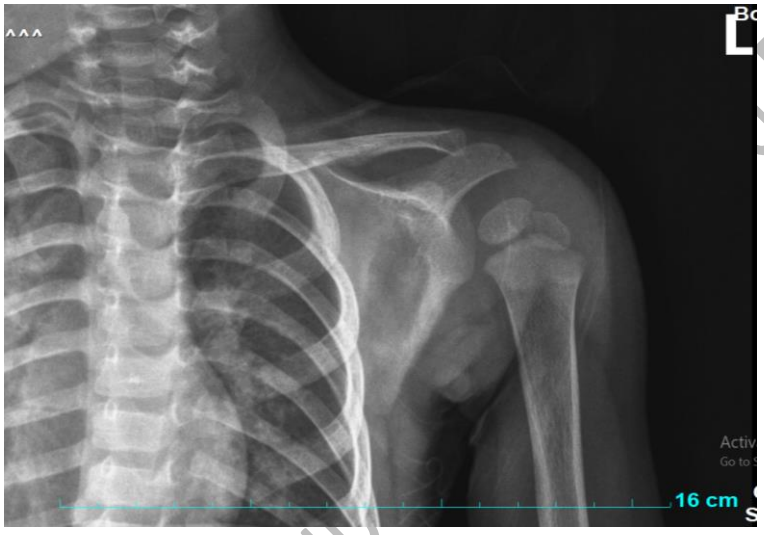


Figure-2

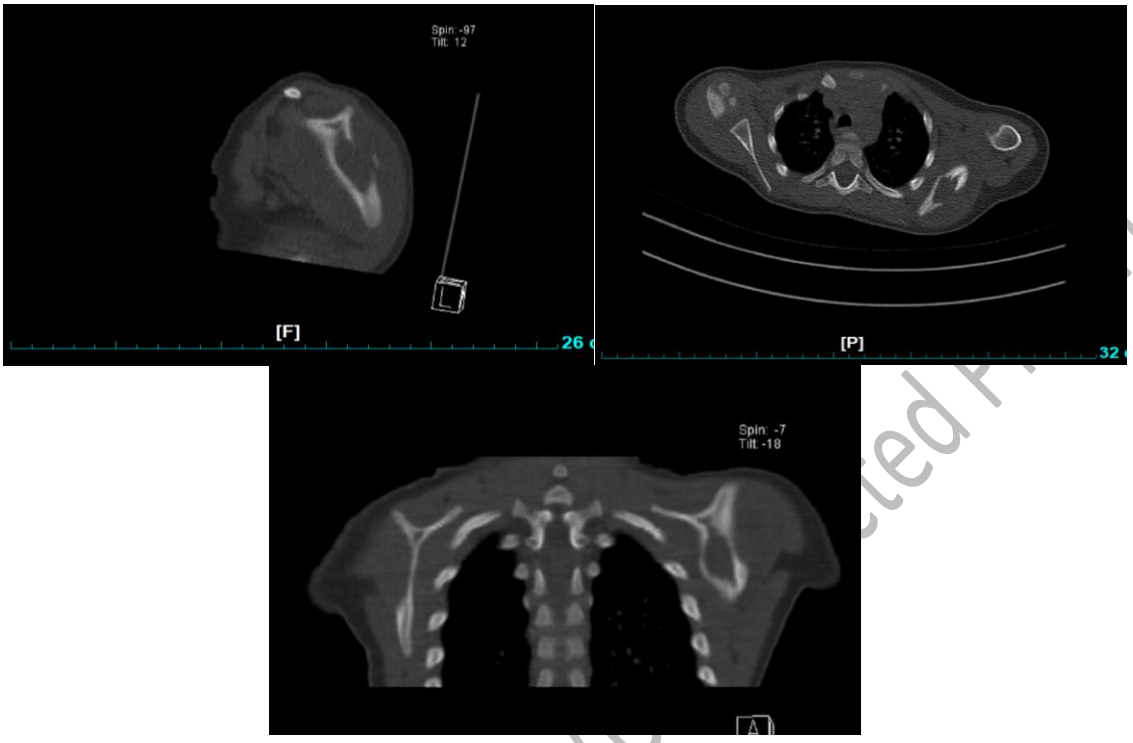


Figure-3

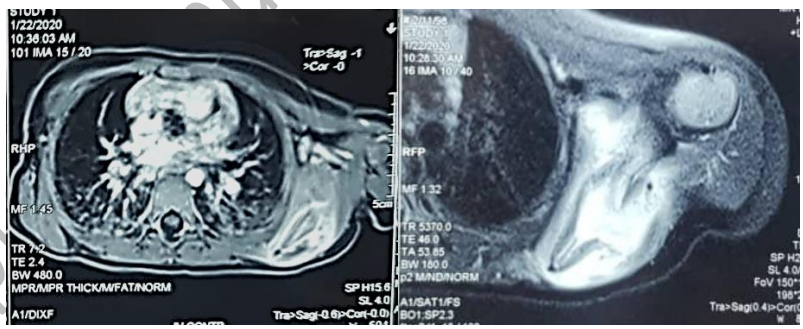
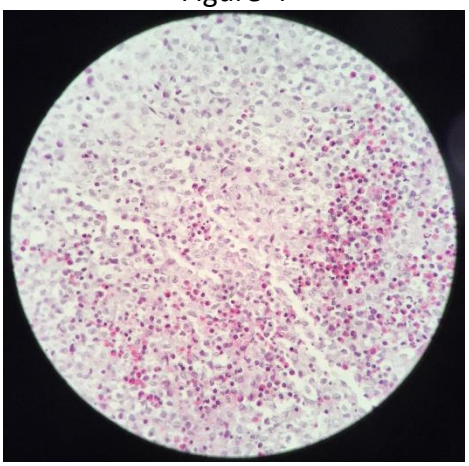


Figure-4



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