Langerhan’s Cell Histiocytosis of Right Scapular Bone: a case report

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Reported here is a case of Langerhan’s cell histiocytosis (LCH), which occurred to a 1-year-old boy presented initially with fever and right-shoulder swelling. In this case, plain radiography and CT revealed an ill-defined osteolytic lesion associated with surrounding soft tissue mass in the right glenoid area. The boy underwent debridement and the specimens were seen to be involved by the Langerhan’s cell microscopically. Vertebra plana was found at L2 vertebra after long bone survey.

Langerhan’s cell histiocytosis (LCH) can occur to individuals of any age, but most commonly to children with a slight male predominance. It was previously called histiocytosis X and defined as an abnormal proliferation of the Langerhan’s cells in various organs or tissues [1]. The reticuloendothelial system (i.e., bones, skin, lymph nodes, liver, and spleen) is involved in most cases, and osseous involvement is the most common manifestation [2]. To our knowledge, a scapular lesion has rarely been found as the initial presentation of LCH. Based on the boy’s clinical history and his presentation at this time, the case may easily be mistaken as a case of osteomyelitis.

CASE REPORT

A 1-year-old boy with fever for one day was taken to our emergency department. His physical examination revealed swelling over his right shoulder joint. The initial hemogram and biochemical analyses showed elevated the white count and ESR. Other biochemical results, including plasma creatinine, liver function tests, and coagulation studies, were within normal ranges.

Plain film of his right shoulder joint showed an ill-defined osteolytic lesion associated with surrounding soft tissue mass in the right glenoid area (Fig. 1).

CT examination of his right shoulder with soft tissue window setting and contrast enhancement revealed a heterogeneous enhancement lesion in the glenoid area of scapula with adjacent soft tissue swelling (Fig. 2a). Bone window setting showed an osteolytic lesion without sclerotic border causing cortical break through (Fig. 2b). Decreased height of the L2 vertebral body, producing the “coin-on-edge” or “vertebra plana,” appearance was also seen in long bone survey. The Tc-99mMDP whole-body bone-scan showed an increased uptake near the glenoid area of right scapular (Fig. 3).
Surgical debridement for the lesion of the shoulder was performed. The histological evaluation revealed that all the tissue fragments contained the Langerhan's cell, which is an unique histiocyte and a distinctive pathologic component of the LCH. Under the diagnosis of LCH with spinal involvement, chemotherapy was given to the boy.

**DISCUSSION**

Langerhan's cell histiocytosis (LCH) is a disorder characterized by the accumulation of the Langerhan's cells. It demonstrates a broad spectrum of clinical and radiologic features. Three major classic syndromes are involved which may have considerable overlapping: 1) Eosinophilic granuloma: usually involving a single bone or a few bones in patients between 5-15 years of age. 2) Hand-Schüller-Christian disease: characterized by multifocal bony lesions and extra-skeletal involvement in children between 1-5 years of age with a chronic recurring course. 3) Letterer-Siwe disease: disseminating involvement of the RES with a fulminant course in patients less than 2 years old [2]. The prognosis of LCH is usually poor in cases with vital organ (hematopoietic, hepatic, or pulmonary) dysfunction. Without vital organ dysfunction, the prognosis would be better [3].

The clinical presentation of LCH depends on the extent of dissemination. Osseous involvement is the most common manifestation of LCH and flat bones including skull, pelvis, and ribs account for more than 50% of all locations of involvement [2].

The bone lesions of LCH typically is osteo-

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**Figure 1.** Plain film of the 1-year-old boy showed an ill-defined osteolytic lesion associated with surrounding soft tissue mass in the right glenoid area (arrow). The vertebra plana is also seen at L2 vertebra (arrow head).  

**Figure 2.** CT examination of his right shoulder joint.  
2a. Soft tissue window with contrast enhancement revealed a heterogeneous enhancement lesion in the glenoid area of scapula with adjacent soft tissue swelling (arrow).  
2b. Bone window setting showed the osteolytic lesion in the glenoid area causing cortical break through (arrow).
lytic, with poorly defined margins and lamellated periosteal reaction in the early phase. During the late phase, the bone lesions appear more well defined and may show sclerotic margins and some degree of expansion and remodeling [2]. Localized osseous LCH is often confused clinically with infection because patients may have a low-grade fever, elevated ESR, mild leukocytosis, as the case we presented here.

The scapula is not a common location for LCH. It only accounts for about 6% of the bone lesions of LCH statistically [2]. Scapular lesions may appear ovoid and osteolytic with or without a circumferential sclerotic border [2]. Serpiginous configuration may be observed sometimes in the scapular lesions [4].

The differential diagnoses of a solitary osteolytic lesion at the scapula with soft tissue mass should include: osteomyelitis, Langerhan’s cell histiocytosis, leukemia, lymphoma, metastasis, Ewing’s sarcoma and rhabdomyosarcoma [5]. Osteomyelitis occurs far more frequently than neoplasms do. Among the neoplasm, metastatic neuroblastoma is the most common cause in children [5].

Vertebral body collapse produces vertebra plana, known as the “coin-on-edge” appearance. The intervertebral disk spaces are always preserved, which help distinguish LCH from pyogenic and non-pyogenic spondylitis [2]. The vertebra may regain almost its original height after many years, with or without treatment [6]. In children, vertebra plana is most often caused by LCH, although it may also be caused by leukemia and metastatic neuroblastoma [2].

Treatment is directed by the clinical situation. More aggressive approaches are for patients with more extensive, multisystem involvement. Solitary bone lesions can be treated with surgical curettage at the time of biopsy when the lesion is readily accessible[7, 8].

In conclusion, Langerhan’s cell histiocytosis with initial clinical presentation of scapular involvement in a 1-year-old boy is unusual. Given the uncommon location of the scapular bone and the clinical presentation of low-grade fever, elevated ESR, and mild leukocytosis, determining the precise etiology is certainly difficult.

REFERENCE

1. Fluri S, Gebbers JO. Langerhans cell histiocytoses: 50 years to histiocytosis X. Schweiz Rundsch Med Prax. 2004; 93: 559-565
朗格漢斯細胞組織細胞增生症發生在右側肩胛骨：
病例報告

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本篇病例報告是關於一位一歲大的男孩以發燒及右邊肩膀腫大的症狀為初始表現。他的胸部X光片及電腦斷層影像顯示了一個界線不清楚的溶骨性病灶及周圍的軟組織腫脹。全身長骨X光片檢查也發現了第2節腰椎骨扁平化的現象。後來病患接受了右邊肩膀病灶處的清創，而病理組織切片證明了朗格漢斯細胞組織細胞增生症的診斷。