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Case report

Ultrasound-assisted diagnosis of Langerhans cell hyperplasia of the sternum: A case report ☆

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ABSTRACT

Langerhans cell hyperplasia is a group of diseases characterized by the proliferation or dissemination of the Langerhans cell, which can come in the form of localized benign lesions, or extensive disseminated invasive neoplastic lesions. These lesions mainly invade the patient's bone. In this paper, we describe the case of a 3-year-old boy who was admitted to the pediatric department with sternum pain. Following a focused sternum ultrasound, a diagnosis was made of Langerhans cell histiocytosis.

Introduction

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by a proliferation of cells that cause local or systemic effects [1]. This mainly occurs in the skull and long bones in the shape of unifocal bone lesions, however multifocal single-system or multi-system forms should be excluded. It is a relatively uncommon disease usually found in infants and children [2]. The involvement of the sternum in LCH is very rare however and few have been observed under ultrasound. It is this rare form of LCH that we report in this observation.

Case representation

A 3-year-old Chinese boy was admitted to our hospital with chest pain in October 2020. Ten days previous to admission, he had developed a pain in the sternum area without any obvious cause, and the pain increased when touched and pushed down upon. He had not shown any obvious discomfort previously, and so he had not been given any special treatment. Three days before admission, the child developed a paroxysmal cough without any obvious cause. He had no fever, no vomiting nor diarrhea. He took oral "amoxicillin granules, Chuanbei sputum cream" for a duration of 3 days and the cough was relieved. However, the pain in his sternum continued to increase. He was apyretic with normal vital signs, had

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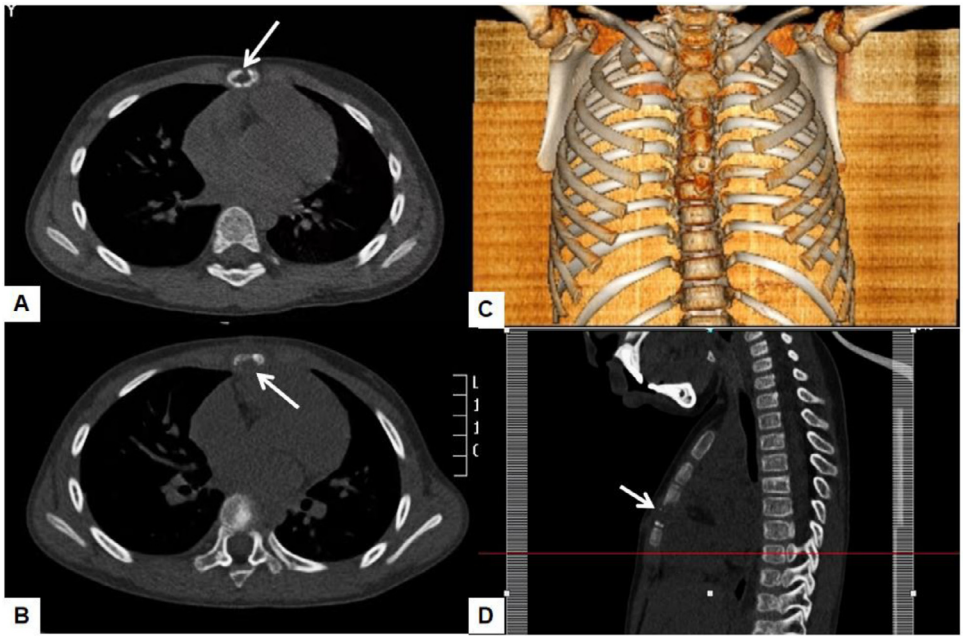

Fig. 1 – Computed tomography findings: (A.B.D) The sternum cortical discontinuity is visible in the area indicated by the arrow. (C) Three-dimensional image reconstruction with fusion of computed tomography imaging.

no fever and had a good general condition. In addition, his routine blood and urine tests were normal.

A computed tomography (CT) of the thorax gave a negative result for pulmonary nodules but showed that the sternum cortical discontinuity was visible and that there was an osteolytic lesion in the sternum. Further examinations were therefore recommended (Figs.1A and B). The osteolytic disease was identified using a 3-dimensional reconstruction and image fusion (Fig. 1C). We therefore performed an ultrasound examination. The ultrasound showed an irregular ill-defined hypoechoic collection overlying the periosteal disruption of the sternum where the lesion was located (Fig. 2). The clinician recommended a needle biopsy. We took the necessary tissue from the lesion using a puncture needle guided by the ultrasound and sent it for pathological analysis (Fig. 2D). A histological examination revealed evident cells showing the typical characteristics of Langerhans cells in some specimens, with some cells found to have the following immunophenotype: S-100 (+), CD68 (+), CD1a (+), and a Ki-67 positive rate of about 20% (Fig. 3). The child then underwent bone marrow puncture and bone marrow cytology, with the results showing: Tissue cells such as large cell bodies, poor differentiation, and an easily visible abnormal shape in the smear. The final pathological analysis confirmed the Langerhans cell hyperplasia diagnosis, and the patient thus underwent regular chemotherapy.

Discussion

Langerhans cell histiocytosis (LCH) is a rare disorder of the myeloid precursor cells, characterized by the uncontrolled proliferation and accumulation of CD1a+/CD207+ dendritic cells [3]. The incidence per annum of Langerhans cell histiocytosis is in approximately 5 people per 1 million, with most cases occurring in children and with a male-to-female ratio of 4:1.1. The tissues and organs that are most frequently affected by Langerhans cell histiocytosis infiltration include the skeleton (80%), skin (33%), and the pituitary system (25%), with the other organs affected, but to a lesser extent, being the liver, spleen, hematopoietic system, lungs (15% each), lymph nodes (5%-10%), and central nervous system excluding the pituitary system (2%-4%) [4]. The single-system, which only involves a single organ with a multifocal or unifocal localization, is usually associated with a good prognosis [5]. The multisystem meanwhile, in which more organs are affected, is associated with a better prognosis when the skin, bones, lymph nodes or pituitary gland are involved, but a worse prognosis when the bone marrow, liver, spleen or lungs are involved [6].

Local bone pain is the most common feature of LCH. Howarth et al. reported that among all the patients with bone lesions that were included in their study, the most frequent osseous invasion sites were the skull (29.9%), proximal femur (12.4%), and ribs (11.1%). Conversely, only 2/314 cases (0.6%) invaded the sternum [7]. As far as we know, only 14 patients with Single System LCH (SS-LCH) of the sternum have ever been reported [8-10]. All these patients successfully recovered without focal recurrence. An LCH diagnosis depends on the patient's history, physical examination, imaging studies, histopathology and immunohistochemistry. Imaging studies are necessary for diagnosis and thus almost all patients underwent an X-ray, CT and Magnetic Resonance Imaging (MRI), however, only a few underwent an ultrasound examination.

The radiographic appearance of bone lesions is that of a medullary lytic lesion with a sharp, scalloped endosteal margin. It is usually non–expansile but apparent expansion may be present due to linear periosteal reaction [11]. The MRI find-

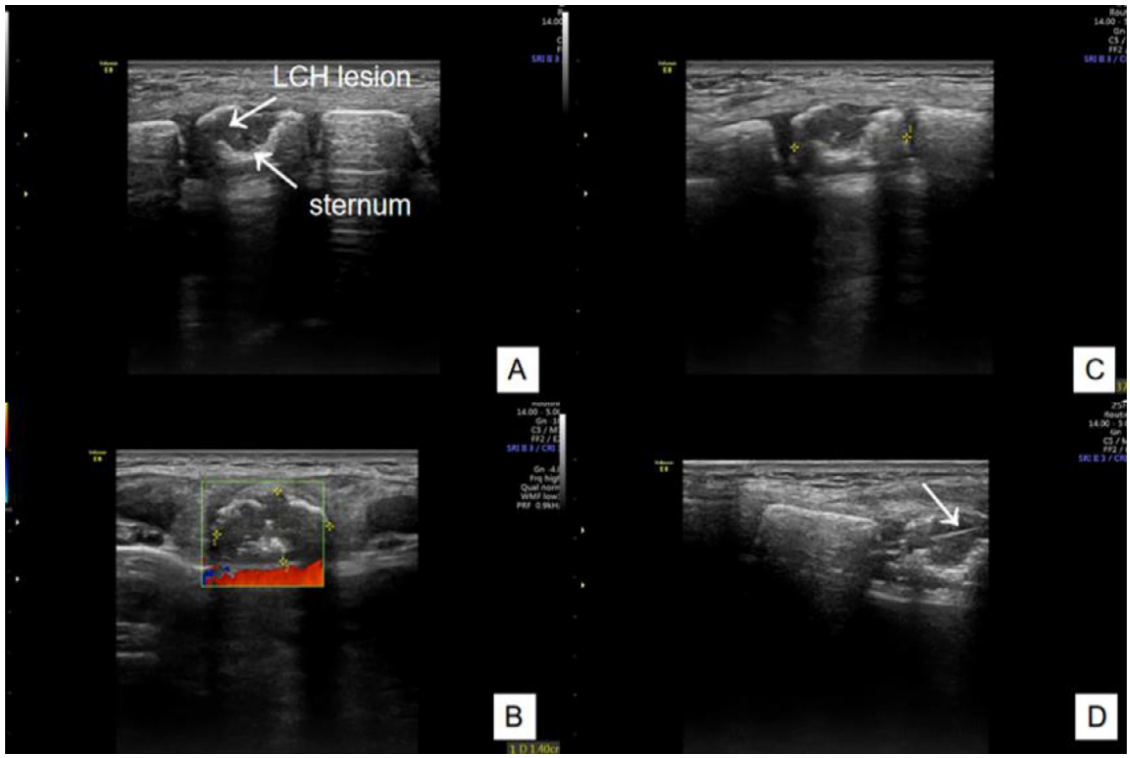

Fig. 2 – Sonogram of the sternum showing a hypoechoic collection overlying the periosteal disruption of the sternum where the Langerhans cell histiocytosis (LCH) lesion is located (A.B.C). Hyperechoic puncture needle inside the sternum lesion (arrow)(D).

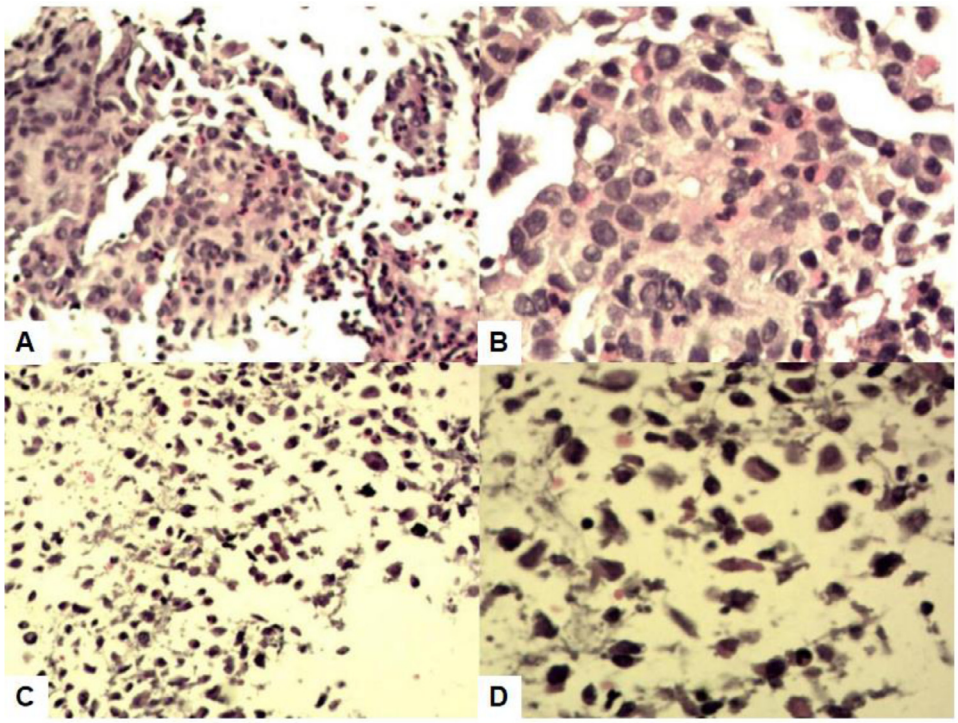

Fig. 3 – A histopathological examination of a sternum tissue biopsy sample. A(10x). B(40x). The hematoxylin and eosin (H&E) stain reveal Langerhans cells with ovoid nuclei and occasional nuclear grooves in a mixed inflammatory background with prominent eosinophilia. C(10x). D(40x). S100-positive and CD1a-positive immunohistochemical staining of Langerhans cells.

ings in LCH of the bone are non–specific. A focal lesion with extensive soft tissue and marrow edema meanwhile is most commonly identified as the hypointense areas in T1Weighted Image (T1WI) images and hyperintense areas on T2Weighted Image (T2WI) and Short T1 Inversion Recovery (STIR) images [12]. In our case, as previously mentioned, the ultrasound examination showed the typical appearance of Langerhans cell histiocytosis sternal lesions, which is a focal and well-demarcated oval or round lesion of very low echogenicity and which shows an evident destruction of the bone. The differential diagnoses include osteomyelitis, osteoblastoma, Ewing's sarcoma, acute leukemia, Gaucher's disease, and metastatic tumor [13]. There are other very useful methods used in these types of diagnoses. For example, ultrasonography has its own unique advantages, including the ability to dynamically observe the extent of sternal invasion in children. Percutaneous radiological guided bone biopsy meanwhile, is now recognized as a safe diagnostic procedure. Furthermore, ultrasound is used in the diagnostic armamentarium of bone diseases and for guiding bone lesions biopsies, as this is a cheaper, radiation-free and widely available imagistic method. In fact, during the ultrasound examination with color Doppler modes, some vascular structures were identified which could then be avoided during the biopsies.

Patient consent

Patient consent has been obtained.

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