초음파 유도하 생검을 통해 진단된 하악에 발생한 Langerhans Cell Histiocytosis

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Langerhans Cell Histiocytosis Arising from the Mandible as Diagnosed by US-guided Core Biopsy

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Langerhans cell histiocytosis (LCH) is a clonal proliferative disorder of Langerhans cells. Although LCH is not considered a malignant disease, its appearance on radiographs may be similar to that of a malignant tumor. The diagnosis of LCH is usually made by a soft tissue biopsy, or by bone marrow aspiration or curettage. We present a patient with a mandibular mass confirmed to be LCH by US-guided core needle biopsy, and present a strategy for diagnosing localized LCH of the bone based on the usefulness and reliability of the percutaneous biopsy.

Key words : Langerhans cell histiocytosis; Mandibular mass; Eosinophillic granuloma on mandible; US-guided core needle biopsy; Bone mass

Introduction

Langerhans cell histiocytosis (LCH) is a clonal proliferative disorder of Langerhans cells [1]. Three disease variants are included in this description: unifocal disease (solitary eosinophilic granuloma), multi-focal unisystem disease (including cases of Hand-Schuller-Christian syndrome), and multi-focal multi-system disease including cases of Letterer-Siwe syndrome [2, 3]. Bone is the most frequently affected organ; the majority of patients present with solitary or multiple bone lesions, and involvement of the jaw is not unusual [4]. Although LCH is not considered a malignant disease, its appearance on radiographs may be similar to that of a malignant tumor [5].

The diagnosis of LCH is usually made by a soft tissue biopsy, or by bone marrow aspiration or curettage. If the lesion is confirmed as LCH with a core biopsy, therapeutic surgical excision is typically planned. For example, if the lesion is revealed to be a malignant bone lesion, a secondary operation or chemotherapy could be needed, while if the lesion is benign, a therapeutic excision biopsy would be planned. To our knowledge, few studies have considered whether ultrasound (US)-guided core biopsy is useful for preoperative pathologic confirmation of LCH.

We present a patient with a mandibular mass confirmed to be LCH by US-guided core needle

biopsy, and present a strategy for diagnosing localized LCH of the bone based on the usefulness and reliability of the percutaneous biopsy.

Case Report

An 18-year-old girl was referred to our institution by her general dental practitioner with a four week history of increasing pain and swelling on the right side of her face. The girl was otherwise healthy with no complicating medical disorders. Upon examination, there was a hard, tender mass that originated from the right ramus of the mandible. All her teeth in the quadrant were vital and non-mobile. Opening of the mouth was slightly reduced and submandibular lymphadenopathy was evident on her right side. The hematological findings were nonspecific.

Contrast enhanced CT images showed an 2.3 cm-sized expanded destructive mass, which had rim-enhancement arising from the ramus of the right mandible and adjacent to the lower right second permanent molar (Fig. 1). The cortex was disrupted and the mass extended into the soft tissues.

A bone scan was performed and there was an abnormal hot uptake lesion at the angle of the mandible with no other skeletal lesions noted (Fig. 2). This lesion was also found to be metabolically active with 18F-FDG-PET (Fig. 3). Our presumptive preoperative clinical diagnosis was a malignant bone tumor including osteosarcoma.

High resolution US imaging performed on the lesion showed a bulging soft tissue mass originating from the

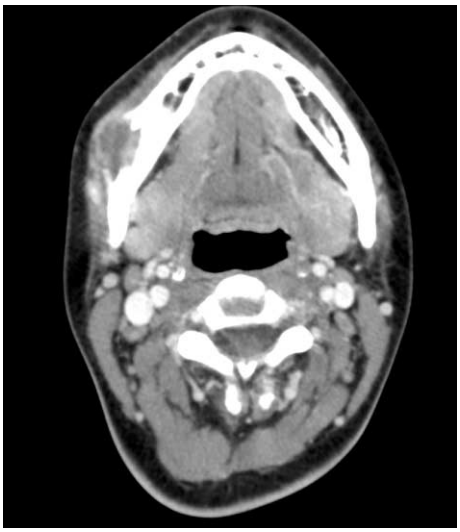

Fig. 1. Axial CT scan shows invasive, destructive borders of the lesion and soft tissue mass on the lateral aspect of the right mandibular body.

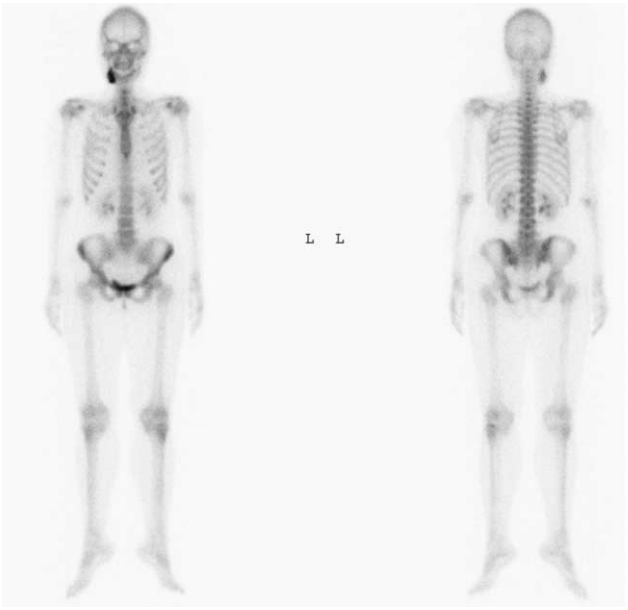

Fig. 2. Whole-body scintigram shows marked radiotracer uptake at the right mandible. There was no uptake in other skeletal system components.

Fig. 3. 18F-FDG-PET shows intense metabolic activities at the right mandible.

right mandible, which had relatively heterogeneous echogenecity with an irregular margin (Fig 4). Color Doppler images showed slightly increased blood flow. In addition, US-guided biopsy was performed by a radiologist, using a 5-12 MHz machine (IU22; Phillps, Bothell Wash) with a 14-gauge multi-fire needle (TSK ACECUT Biopsy System,Tochigi, Japan) under local anesthesia; 4-core specimens (needle excursion length; 2 cm) were obtained. No specific complication was noted and the lesion was revealed to be LCH (Fig. 5). The planned surgical excision was performed and the diagnosis was the same. The patient has had no evidence of active LCH during one-year of follow-up.

Discussion

LCH is a reactive disorder characterized by the infiltration and proliferation of dendritic cells with the appearance of normal Langerhans' cells [6]. Osseous involvement is the most common manifestation of LCH, occurring in 60-95% of pediatric patients. Solitary bone lesions are typically treated surgically, but medical treatment is indicated for multiple or progressive lesions or bone lesions not amenable to excision [7]. Thus, imaging is used to identify multifocal disease or to give an indication of lesional metabolic activity. Plain radiographs, CT, and bone scintigraphy have been the traditional modalities used in imaging. However, recent studies have found that bone lesions with high concentrations of histiocytes show increased metabolic activity with FDG-PET.

The radiologic appearance of osseous LCH depends on the site of involvement. Lesions typically appear as 'expandable' osteolytic but may have either poorly defined borders or well-demarcated margins, and with or without reactive sclerosis. CT imaging is useful in providing detailed cross-sectional anatomic details of the involved bone. However, these findings are nonspecific compared to the findings of other soft tissue masses arising in the skeletal system. US imaging is also limited for the description of a soft tissue mass, which can just demonstrate the space-occupying mass within bone, with or without a cortical disruption.

Because LCH lacks pathognomonic clinical or radiographic characteristics, a definitive diagnosis should be based on the histologic and immunohistochemical study of lesional biopsy specimens. Radiographically, the differential diagnosis includes osteomyelitis and Ewing's sarcoma in children and metastatic carcinoma as well as other benign and malignant primary tumors of bone in adults.

These soft tissue and bone masses that cannot be

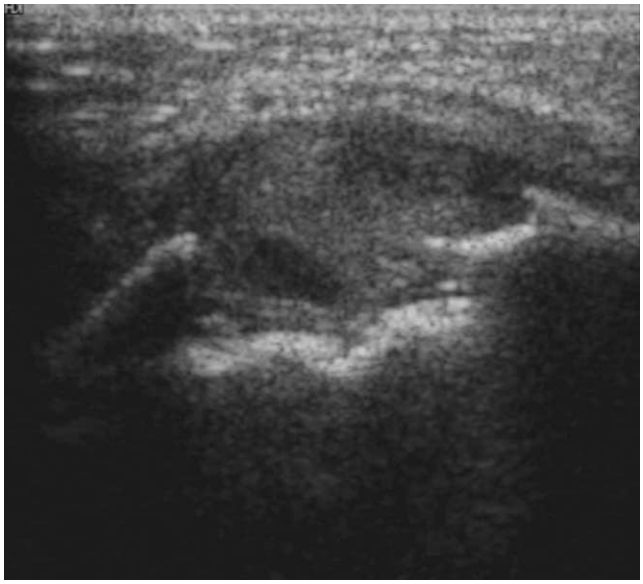

Fig. 4. Ultrasonograms of the mandible show a soft tissue mass arising from cortex of the mandible with cortical destruction.

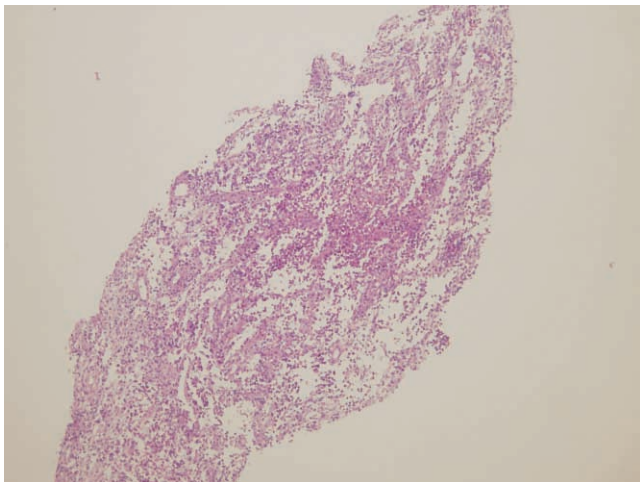

Fig. 5. The core biopsy specimen shows diffuse infiltration of Langerhans cells admixed with inflammatory cells, including eosinophils. The diagnosis was Langerhans cell histiocytosis (H & E, ×40).

identified by imaging studies are considered indeterminate and a surgical or percutaneous biopsy is usually necessary to identify them [8]. An open biopsy has been considered as the gold standard for the initial diagnosis of soft tissue and bone masses. While an open biopsy can fetch a large amount of tissue, general anaesthesia is necessary, which increases the cost of the procedure [9]. In addition, up to 50% of microscopic residual disease has been reported in sarcomas when an excisional biopsy (enucleation) was performed [10, 11]. A percutaneous core needle biopsy is a minimally invasive, inexpensive, and efficient diagnostic method which is increasingly used in the initial approach to these types of tumors, with a reported diagnostic accuracy ranging from 78% to 97% [12, 13]. Although percutaneous needle biopsy may result in sampling errors because of the small amounts of tissue obtained, the combination of guidance from CT or US, reliable immunohistochemistry, and experienced pathologists can show a higher sensitivity and specificity rates [13−15].

Image guidance is generally required for biopsies of head and neck lesions. CT, with its high spatial and contrast resolution, is the imaging modality of choice for biopsies of deep-seated head and neck lesions. CT allows excellent delineation of intervening vital structures, which permits a safe biopsy path planning. With CT, the operator can target the appropriate part of the mass and avoid areas of necrosis. On the other hand, US-guided core needle biopsy, with its inherent advantages of real-time imaging capability, decreased procedural time, ability to visualize vessels without use of intravenous contrast material, portability, lack of ionizing radiation, and decreased cost, allows for the confirmation of the superficial-seated head and neck lesion as LCH, and avoids unnecessary excessive surgery. Additionally, by high-resolution US imaging of superficial skin lesions, the anatomy can be defined, lesions characterized, and blood flow confirmed, all without the use of ionizing radiation [7]. In our case, US imaging of the mass on the right mandible provided tumor characteristics, disease extent, as well as guidance for the core biopsy.

However, US examination is limited by the inability of sound waves to penetrate overlying bone [7]. Therefore, US-guided biopsy also could be limited by a soft tissue mass bulging out from the bony cortex, which would require a careful approach during the progression of the needle tip. Major complications from image-guided core biopsies are rare. Minor complications include pain, vasovagal reaction, minor infection and bleeding.

In summary, our observation suggests that a US-guided core biopsy for LCH as a solitary bony mass provides an accurate histologic confirmation and optimal guidelines for a treatment plan.

요 약

랑거한스세포 조직구증 (Langerhans cell histiocytosis, LCH)는 랑거한스세포의 이상 증식을 특징으로 하는 원인 불명의 질환이다. LCH가 악성질환으로 간주되지는 않으나, 영상의학적 형태는 악성종양의 그것과 유사할 수 있다. LCH의 진단은 대부분 연부조직 생검, 골수 흡입생검 또는 소파술로 이루어진다. 우리는 초음파 유도하 조직생검을 통해 LCH로 진단된 하악종괴를 가진 환자에 대한 1예를 보고하고, 골부에 국한된 LCH를 진단할 수 있는 경피 조직검사의 유용성과 안정성을 보여주고자 한다.

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