Case Report

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Langerhans Cell Histiocytosis in the Nasal Bone: A Rare Case

코뼈에 발생한 랑게르한스 세포 조직구증: 증례 보고

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Nasal bone involvement of Langerhans cell histiocytosis is rarely reported. Here we present a case of a 13-year-old boy with a palpable nasal mass. Ultrasonography revealed a hypoechoic mass on the left side of the nose. Both CT scanning and MRI showed an osteolytic mass. The lesion seen on MRI was well-defined mass with homogeneous enhancement. Histopathological examination of the resected specimen confirmed the diagnosis of LCH.

Index terms Langerhans Cell Histiocytosis; Nasal Bone; Ultrasonography; Computed Tomography, X-Ray; Magnetic Resonance Imaging

INTRODUCTION

Langerhans cell histiocytosis (LCH) is an uncommon condition involving clonal proliferation of Langerhans cells. The reported incidence of LCH ranges from 0.2–0.5 cases per 100000 children per year (1-3). LCH predominantly affects children aged 5–15 years (3, 4) and is more prevalent in males than females (M:F; 2:1) (3, 5, 6). Although its etiology remains unknown, uncontrolled proliferation and accumulation of bone marrow-derived immature myeloid dendritic cells are presumed to play a role (1, 6, 7).

LCH may manifest as a unisystem (unifocal or multifocal) or multisystem disease (1).

Although there have been several radiologic reports of LCH affecting the head and neck, such as the petrous temporal bone, sphenoid bone, and orbits, involvement of the nasal bone is very rare (1, 5, 6). Herein, we report a case of LCH affecting the nasal bone.

CASE REPORT

A 13-year-old boy presented with a 3-day history of a palpable mass on the left side of his nose. There were no other nasal symptoms. The patient's family history, medical history, and laboratory findings were unremarkable. On physical examination, there was tenderness around the site of involvement.

Ultrasonography revealed a hypoechoic mass on the left side of the nasal dorsum (Fig. 1A). The mass involving the nasal bone was seen to extend into the subcutaneous layer. CT scanning of the facial bone revealed a 1.8 cm × 1.2 cm osteolytic soft tissue mass with lateral protrusion along the anterior margin of the left maxilla (Fig. 1B, C). MRI revealed a well-defined osteolytic mass (low signal on T1-weighted and high signal on T2-weighted sequences). Contrast-enhanced T1-weighted image showed homogeneous enhancement (Fig. 1D, E).

Excisional biopsy was performed for histopathological confirmation, which revealed proliferation of Langerhans cells with prominent nuclear grooves and admixed eosinophils, and immunohistochemical staining was positive for CD1a and S100. Therefore, the patient was diagnosed with LCH involving the nasal bone. A PET/CT was conducted to screen for other suspicious body parts, which was negative. The patient was treated with and responded well to chemotherapy with vinblastine, and there was no tumor recurrence.

The proceedings in this case were approved by the Institutional Review Board of our institution (IRB No. DFE21ORIO113).

Fig. 1. A 13-year-old boy with Langerhans cell histiocytosis on the left nasal bone.
A. Ultrasonography image shows a hypoechoic mass (asterisk) in the subcutaneous fat layer of the left side of the nose.
B, C. CT image reveals an irregular hypoattenuated soft tissue mass measuring 1.8 cm × 1.2 cm infiltrating the left nasal bone. A focal osteolytic lesion (arrows) with lateral protrusion along the anterior margin of the left maxilla is visible.
D, E. Axial fat-saturated T2-weighted magnetic resonance image shows a well-defined hyperintense mass (arrows). Axial contrast-enhanced T1-weighted image shows a well-defined focal osteolytic mass with diffuse enhancement.

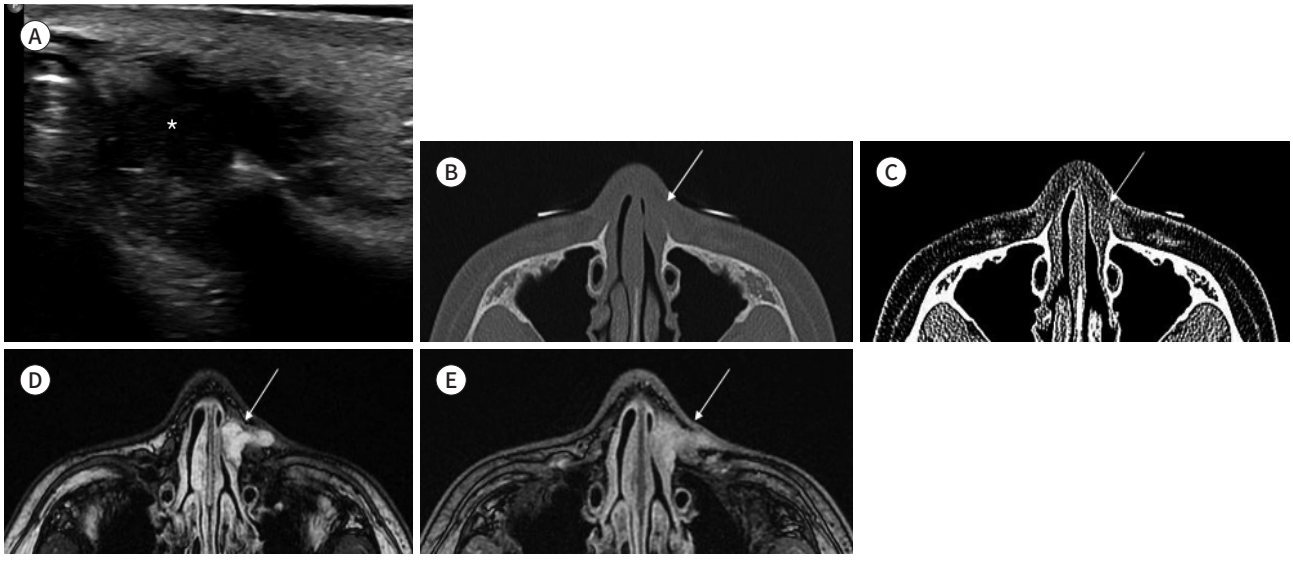
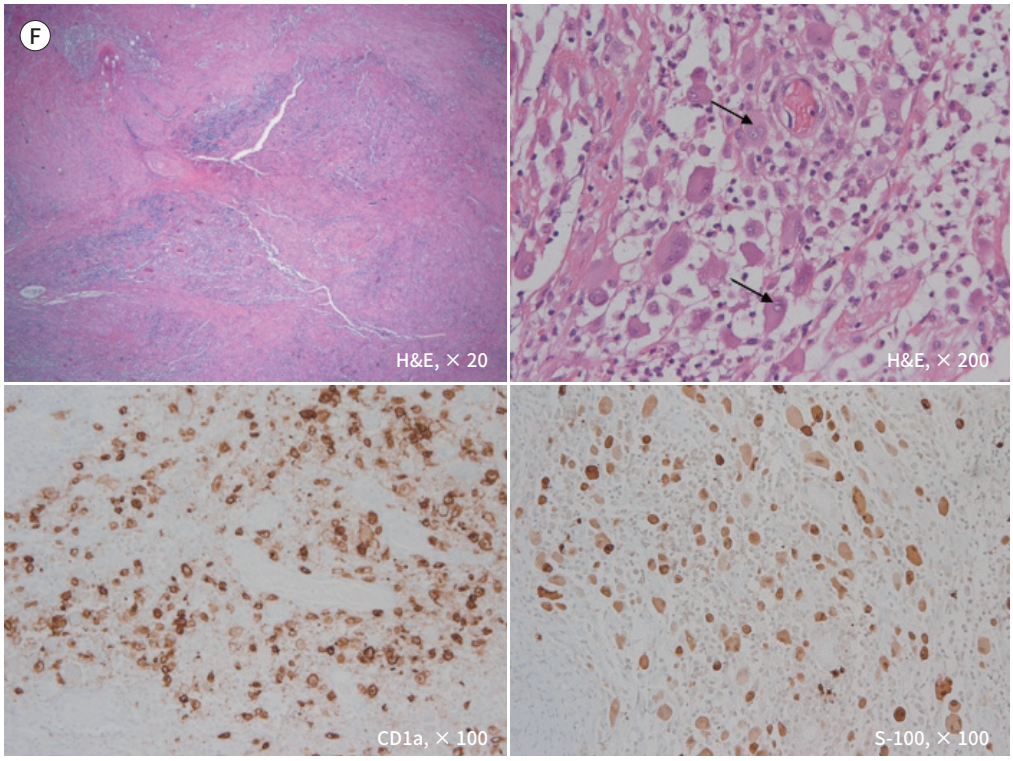


Fig. 1. A 13-year-old boy with Langerhans cell histiocytosis on the left nasal bone. (Continued)
F. Histopathological micrography image of H&E-stained tissue shows mononuclear histiocytes, and acute inflammatory cells, a multinuclear Langerhans cell with prominent nuclear grooves (arrows), and admixed eosinophils. The cells are positive for CD1a and S100 on immunohistochemistry.
H&E = hematoxylin and eosin



DISCUSSION

LCH is an inflammatory myeloid neoplasia characterized by the accumulation of clonal histiocytes and granuloma formation in affected tissues. Despite its unknown etiology, uncontrolled proliferation and accumulation of immature myeloid dendritic cells derived from bone marrow is thought to be the cause (1). The most common symptom is localized pain (48.4%) followed by swelling (26.5%). A diverse range of symptoms occur due to the involvement of other body parts (3, 4).

The incidence of infant LCH is 0.2–0.5 in 100000 (3). Although LCH involving bony structures is more common, LCH can involve any organs despite single site involvement being more common than multi-site involvement (3, 4). Although LCH has been reported to affect the head and neck, involvement of the nasal bone is rare (6, 7).

The most common radiological feature of LCH is an osteolytic lesion that may present as a moth-eaten, punched-out, geographic, or expansile form (8). On CT, the soft tissue component of the tumor appears homogeneous and isodense to the muscles both on contrast and non-contrast-enhanced images. On MRI, the soft tissue component appears heterogeneous with a variable pattern on T1-weighted images, while on T2-weighted and short tau inversion recovery (STIR) sequences, it shows heterogeneous enhancement with contrast administration. The images in our case depict a similar pattern. While CT imaging showed a soft tissue mass involving

the nasal bone, MRI revealed hyperintensity on T2-weighted images and hypointensity on T1-weighted images with strong contrast enhancement. The mass assumed a "V" shape on contrast-enhanced MRI, possibly due to its growth toward the soft tissue. As imaging features are non-specific, histopathological examination and immunohistochemical staining (with CD1a, S100 protein, and/or CD207 antibodies) of tissue samples are necessary to confirm LCH (8).

In cases of presenting with solitary osteolytic lesions, several conditions are considered in the differential diagnosis of which epidermoid or dermoid cysts are one. However, epidermoid or dermoid cysts show poor enhancement. In addition, chondroblastoma, which is relatively common in the sphenoid and ethmoid bones, can present similarly. On CT, chondroblastoma appears as an osteolytic lesion with internal calcification. Such calcifications or new bone formation show signal voids on MRI T2-imaging, but depict heterogeneous enhancement and peritumoral edema on contrast administration. Though fibrous dysplasia may appear as osteolytic lesions, it usually appears as ground glass appearance with bone expansion with intact cortical bone. Lymphoma may also present as lytic lesions with a non-specific enhancement pattern, but it is uncommon in children (9).

The treatment and outcome of LCH depend on the extent and severity of the disease. Unifocal bone involvement is usually a mild form of the disease, which is treated locally by excision, curettage, intralesional steroid injection, or radiation therapy (8). The outcome of such treatment is good, with more than 80% of patients recovering completely (10). However, multifocal bone involvement or multisystem disease affecting the bones requires systemic or combination therapies, and their prognosis depends on the site of the bone lesion and the organs involved. Multisystem involvement with "risk organs" such as the liver, spleen, and bone marrow is associated with a poor prognosis (8).

In conclusion, we report a rare case of LCH involving the nasal bone. Differential diagnosis of LCH from other bone tumor is difficult even for experienced radiologist. However, in cases of craniofacial osteolytic lesion in children, LCH should be considered one of the possible causes, which may allow proper diagnosis and therapy.

Author Contributions

Conceptualization, J.J.C.; data curation, J.J.C.; investigation, J.J.C., A.W.I.; project administration, J.J.C., A.W.I.; supervision, J.J.C., Y.D.J.; validation, J.J.C., Y.D.J.; writing—original draft, J.J.C., A.W.I., Y.D.J.; and writing—review & editing, A.W.I., Y.D.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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코뼈에 발생한 랑게르한스 세포 조직구증: 증례 보고

안원익[1] · 장종창[1]* · 양동진[1] · 김태은[1] · 박효천[2] · 이동규[2] · 성재민[2]

코뼈에 발생하는 랑게르한스 세포 조직구증 증례는 거의 보고된 적이 없다. 저자들은 왼쪽 코에 만져지는 종괴를 주소로 내원한 13세 환자를 검사하였다. 초음파 검사상 코 좌측에 저음영 에코를 보이는 종괴가 관찰되었다. 자기공명영상 검사에서 경계가 분명하면서 비교적 균질한 조영증강을 보이는 종괴가 관찰되었다. 조직병리학적 검사에서 검체는 랑게르한스 세포 조직구증으로 확인되었다.

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