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CASE REPORT



# Reconstruction of a severe mandibular pathological fracture caused by Langerhans cell histiocytosis using a free fibula osteocutaneous flap: a case report

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**ABSTRACT**

Langerhans cell histiocytosis (LCH) is a rare disease in which Langerhans cells, which are bone marrow-derived antigen-presenting cells, proliferate in single or multiple organs. We successfully treated a patient with unifocal LCH of the mandible with malocclusion due to a severe pathological fracture, using reconstruction with a vascularised free bone.

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**KEYWORDS**
Langerhans cell histiocytosis; pathological fracture; vascularised free bone

## Introduction

Langerhans cell histiocytosis (LCH) is a rare disease involving the proliferation of Langerhans cells, the antigen-presenting cells derived from the bone marrow in organs, such as the skin, bone, lymph nodes, lung, liver, spleen and the central nervous system [1]. The disease is most likely to occur in the jaw bone in the oral and maxillofacial areas, and the prognosis is relatively good [2]. We report a case of LCH with a severe pathological fracture of mandibular bone that was reconstructed using a free fibula flap.

## Patient

A 55-year-old man reported pain in the right lower jaw that had started one year previously. Because the pain increased gradually, he was referred to an oral surgeon. Gross examination revealed perforation of an ulcer in the gum on the bottom right of the jaw (Figure 1). An X-ray showed a bone transmission image in the lower right jaw, floating teeth and a severe pathological fracture of the mandibular body (Figure 2). A computed tomography scan showed a cystic lesion on the same site as the pathological fracture (Figure 2). Because occlusal disharmony due to pathological fracture was observed, an occlusal correction using rubber traction was performed. A magnetic resonance imaging (MRI) scan showed a low signal on T1-weighted images, and a high signal on T2-weighted images (Figure 3). Haematoxylin–eosin stain of the biopsy showed infiltration of eosinophils, with Langerhans cell-like cells in the periphery. Immunohistochemical staining was S100 protein positive and CD1a positive (Figure 4); therefore, the diagnosis was LCH. There were no abnormal findings in fluorodeoxyglucose-positron emission tomography (FDG-PET) or MRI scans of the head. Therefore, the diagnosis was LCH confined to the mandible, and a mandibular segmental resection and reconstruction using a free fibula flap was performed (Figure 5).

One month after the operation, an intermaxillary fixation was performed to obtain normal occlusion. Normal occlusion was acquired, and no recurrence was observed more than two years after surgery (Figure 6).

## Discussion

LCH is a generic term that includes eosinophilic granuloma of the bone, Hand–Schüller–Christian disease, Letterer–Siwe disease, which was previously called Histiocytosis X, pulmonary LCH and Hashimoto–Pritzker disease. Although there are

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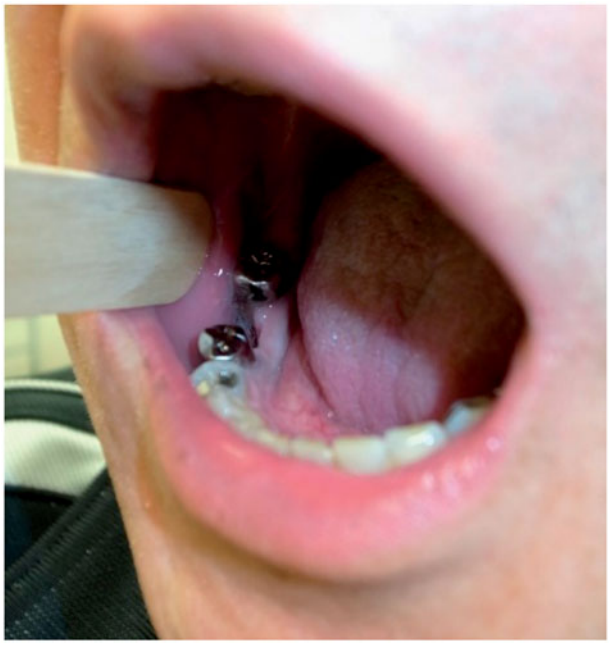

Figure 1. A perforation of the ulcer was observed in the gum on the bottom right jaw.

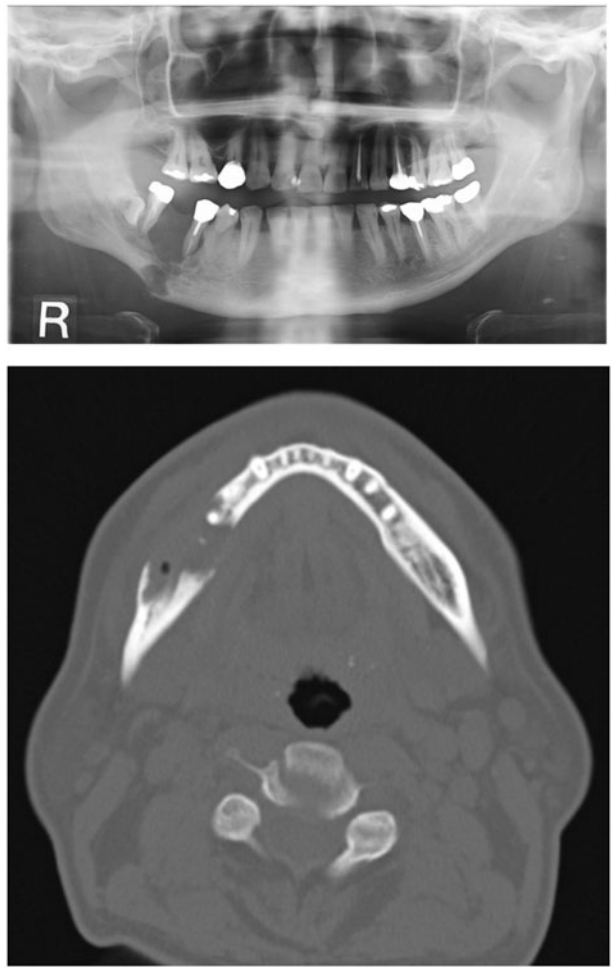


Figure 2. Radiograph showing the bone transmission of the lower right jaw, floating teeth, and a pathological fracture of the mandible body. CT scan shows a cystic lesion on the same site as the pathological fracture.

differences in the peak ages of onset, the locations of the disease and the prognosis, they are all the same disease pathologically [3]. The pathogenesis of this disease is still unknown. The incidence is low and has been reported to be one in 200,000 people [1]. In the Histiocyte Society of the World Health Organisation, LCH is classified into three types: unifocal disease, multifocal single system disease and multifocal multisystem disease [3,4]. The frequency of each is almost the same, ranging from 31% to 36%. Multifocal single system disease and multifocal multisystem disease occur in younger people with increased recurrence rates and mortality [3]. It is reported that approximately 30% of all LCH occurs in the oral and maxillofacial area, commonly in the jaw bone [5] and more commonly in the mandible than the maxilla [2].

LCH of the jaw bone is frequently present and appears as radiolucent single or multiple lesions. These lesions may cause the teeth to appear as if they are floating in space [6]. In MRI, the lytic lesions seen on radiography and CT are low signal, or isointense to muscle, on T1-weighted images and high signal on T2-weighted images [7]. A technetium 99 m-methylene diphosphonate (Tc-99 m MDP) bone scan shows an abnormal radionuclide concentration in the margins of the lesion. Tc-99 m MDP has been particularly useful for the diagnosis of multiple bone lesions [7]. In recent years, FDG-PET has been reported to be useful for a number of diagnoses [8–10].

For a definitive diagnosis of LCH, the following are required: abundant eosinophilic cytoplasm, a large number of Langerhans cell with a kidney-like or deep groove in the nucleus, an associated infiltration of eosinophils, positive reaction to CD1a or Langerin, and the presence of Birbeck granules (which appear as rod-like or tennis racket-like structures in the Langerhans cells) must be confirmed by electron microscopy.

Treatment has not yet been established, but it differs for the single- and multiple-organ types. For the former, surgical curettage and resection, low-dose radiation therapy, and topical administration of steroids are recommended. In addition, LCH may resolve spontaneously, and many reports have been made that it is good to follow up after a biopsy [7]. For example, a case of LCH in the jaw bone began to shrink at 2 to 3 weeks after biopsy, and there are reports that natural healing has occurred within 2 to 36 months [11,12]. For the latter, chemotherapy with vinblastine, cyclophosphamide, cyclosporine or methotrexate, in combination with systemic or topical administration of

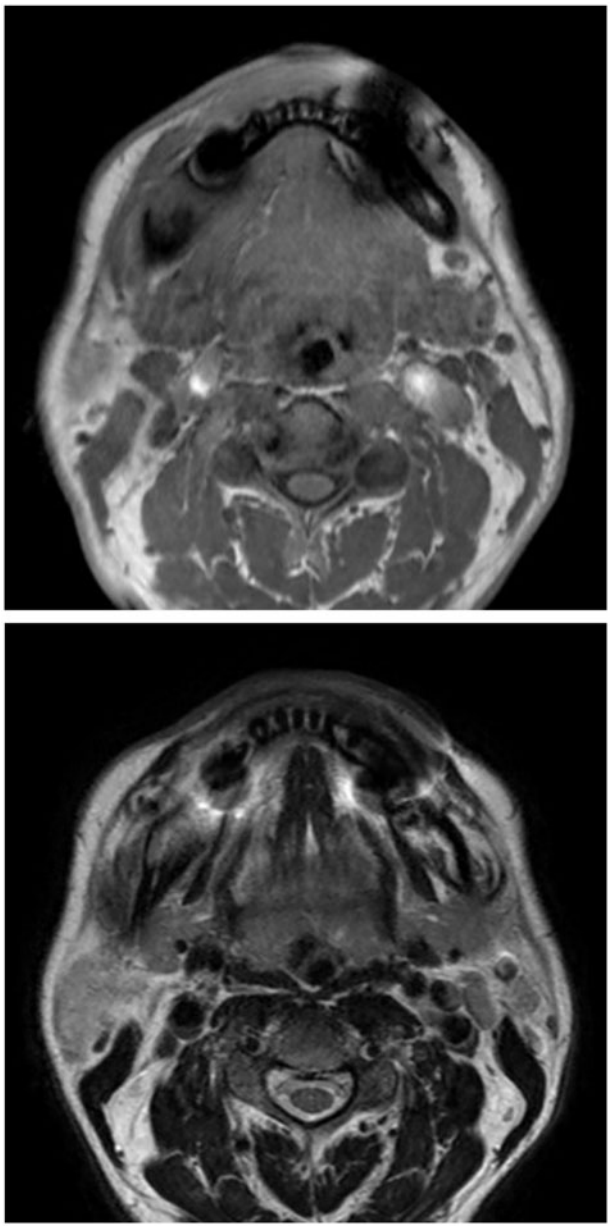

Figure 3. MRI scan shows low signal on T1-weighted images and high signal on T2-weighted images.

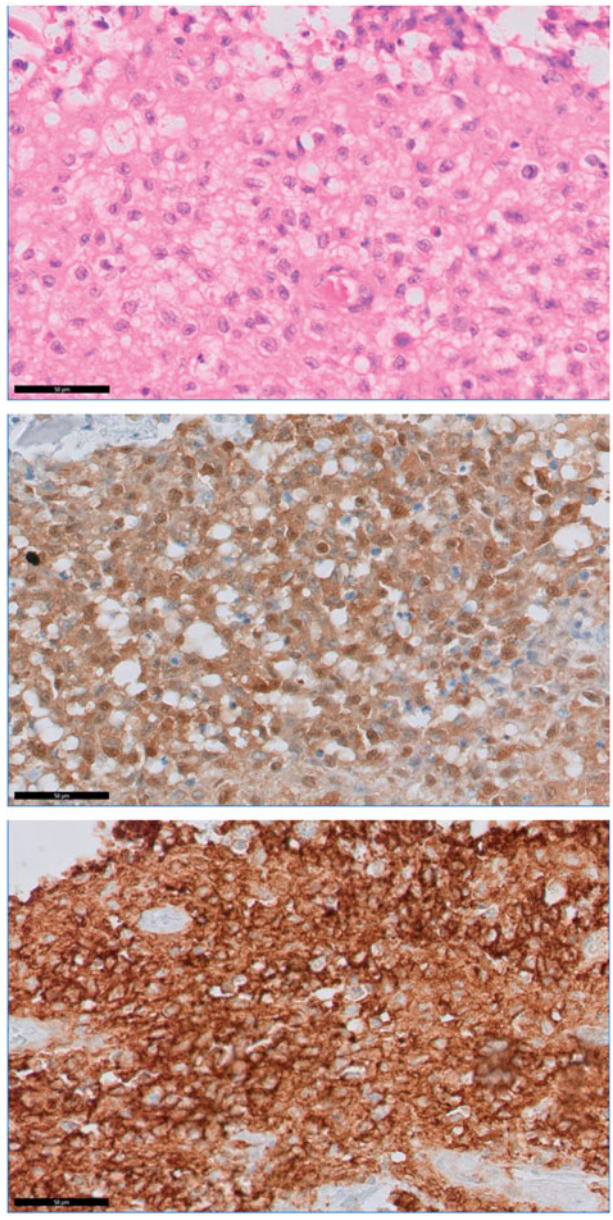

Figure 4. Haematoxylin–eosin staining indicating infiltration of eosinophils. The Langerhans cell-like cells in the periphery were S100 protein positive. The Langerhans cell-like cells in the periphery were CD1a positive.

steroids, has been reported many times, but there are many options for an effective treatment schedule and the combination of drugs [3,7]. There are reports that bone marrow transplant has been effective in severe cases of refractory LCH [13]. In the case of a single-organ type occurring in a single jaw bone, there have been many cases of good healing achieved by surgical curettage and removal, and there are varying opinions as to whether surgical treatment is most recommended [12]. Similar to this case, there are reports of resected lesions that developed in the jawbone and were reconstructed with a free fibular flap [14,15]. On the other hand, there are reports that surgery should be used only when drug therapy fails, or when important tissues such as the optic nerve or spinal cord are affected [16]. There were nine reports of local injections of steroids that were effective [17–23], and the average time to healing was 9.8 months. In this case, based on the fact that it was a single-lesion type, that there was no tendency for shrinkage of the lesion after the biopsy, that the patient was not elderly, that his general condition was good and especially because it was necessary to correct the malocclusion due to a severe pathological fracture as early as possible, segmental resection of the mandible and reconstruction with a free vascularised fibula flap was chosen. As a

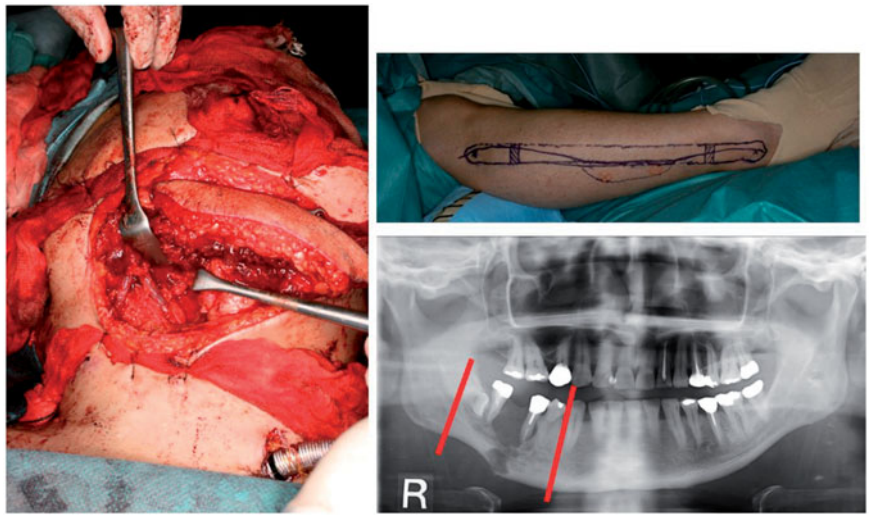


Figure 5. Mandible segmental resection and reconstruction by a free fibula flap were performed.

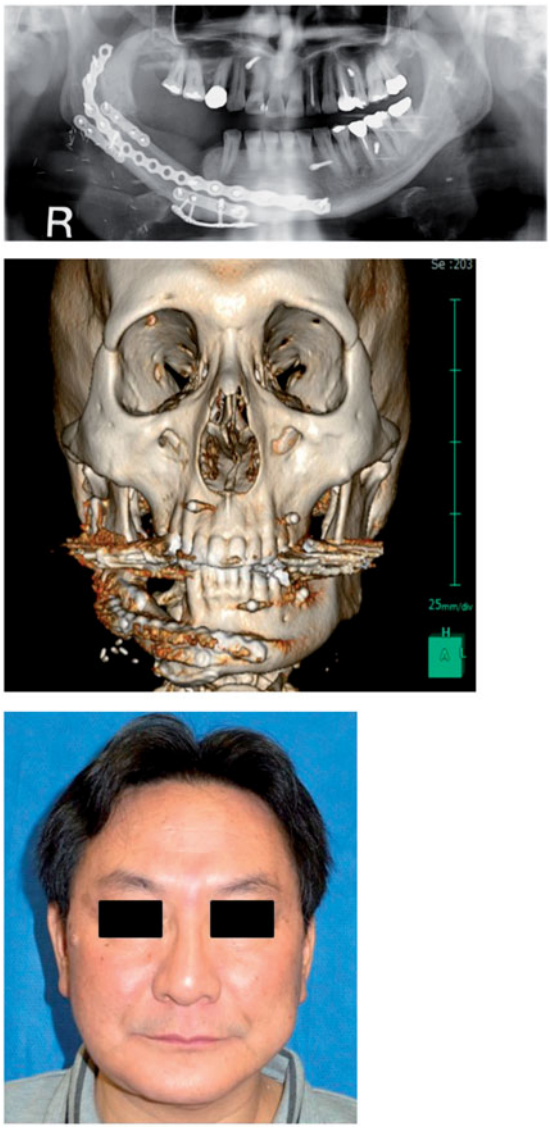


Figure 6. One month after the operation, intermaxillary fixation was performed to obtain a normal occlusion. No recurrence was observed more than two years after surgery.

result, normal occlusion was obtained within a relatively short period of 1 month after surgery.

## Conclusions

For unifocal LCH of the mandible bone with malocclusion due to a severe pathological fracture, reconstruction with a vascularised free bone can be a useful option for treatment to obtain normal occlusion as early as possible.

## Disclosure statement

No potential conflict of interest was reported by the authors.

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