CASE REPORT

Eosinophilic granuloma of the mandible: a diagnostic dilemma

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SUMMARY

Eosinophilic granuloma (EG) is a rare histiocytic disorder resulting from clonal proliferation of Langerhans cells. It accounts for less than 1% of all osseous neoplasms and has a predilection for involving the axial skeleton. Although suspicion of the disease may arise from clinical features and radiographic demonstration of destructive bone lesions, it is still difficult to make a correct diagnosis without proper pathological evaluation. This is more evident when common differentials mimicking EG, both clinically and radiologically, need to be ruled out. This report describes a case of unifocal EG of the mandible occurring in a 4-year-old boy whose initial presentation led to confusion between osteomyelitis, primary bone tumour and lymphoma. A final diagnosis of EG was established after histopathological examination of the biopsy specimen.

BACKGROUND

Langerhans cell histiocytosis (LCH) refers to a relatively rare condition resulting from neoplastic proliferation of Langerhans cells. It is a disease of unknown aetiology and diverse manifestations. Eosinophilic granuloma (EG)—a term used synonymously with LCH by the World Health Organisation—is a localised form of the disease. It is in fact the mildest form of the histiocytosis-X group of diseases, which also encompass Hand–Schuller–Christian disease and Letterer–Siwe disease. The above grouping was based on the similarities of the histopathological appearance of the histiocytic and eosinophilic proliferation.

EG accounts for less than 1% of all osseous neoplasms and occurs more commonly as solitary rather than multiple lesions. The disease has a predilection for involving the axial skeleton, and the incidence is higher among men. More than half of the patients are less than 10 years of age at the time of presentation, although people of any age can suffer from this disorder.

In this report, we present a rare case of unifocal EG of the mandible occurring in a 4-year-old boy who was admitted with complaints of a gradually increasing swelling on the right side of his face over 1.5 years, accompanied by non-healing ulcerative lesions in the lower molar gingiva. Radiological investigations showed the presence of a lytic lesion in the mandible with floating teeth. Based on these findings, the differentials that were considered were osteomyelitis, primary bone tumour and lymphoma. However, biopsy from the lesion showed the characteristic features of EG. We highlight this unusual case and emphasise the importance of histopathological examination in the diagnosis of this rare condition.

CASE PRESENTATION

A 4-year-old male patient was referred to the outpatient department of our hospital with complaints of pain and progressively increasing swelling on the right side of the lower jaw, over the past 1.5 years. There was difficulty in mastication and dysphagia. A low grade fever was associated from the time of onset of the swelling; however, weight loss and a history of trauma were absent. During this entire period, the patient had several trials of antimicrobials but without any effect.

Clinical examination revealed a single large globular swelling measuring approximately 5×4 cm over the right lower jaw region with extension towards the angle of the mandible (figure 1). On palpation, the swelling had diffuse margins, was tender, firm to hard, immobile and attached to the underlying structures. Intraorally, an ulceroproliferative growth was detected around the lower gingiva on the same side (figure 2). Multiple loose teeth were also present.
around the lesion. No regional lymphadenopathy or hepatosplenomegaly was observed.

INVESTIGATIONS
On routine workup, the haemogram was within normal limits except for raised erythrocyte sedimentation rate (32 mm/h). Intraoral periapical radiographs and orthopantomograms showed a radiolucent area in the mandible producing the appearance of teeth ‘floating in air’. CT scan, done subsequently, revealed a lytic lesion extending into the alveolar part of the mandible with intraoral soft tissue extension (yellow arrows). The patient was admitted for further evaluation.

Fine needle aspiration cytology of the lesion remained inconclusive as it showed only mixed inflammatory cells and macrophages. Hence an excisional biopsy was performed and the soft tissue, bony chips and teeth were sent for histopathological evaluation. On gross examination, small fragments of greyish-brown soft tissue along with bony chips and teeth were observed (figure 4). Microscopic examination revealed ulceration of the mucosa accompanied by extensive aggregates of histiocytes showing a reniform nucleus, nuclear grooves and eosinophilic cytoplasm, extending deep into the underlying submucosal tissue (figure 5). Numerous eosinophils were seen lying in clusters as well as individually dispersed. In addition to these, some lymphoid cells, plasma cells and neutrophils were also present (figure 6). No malignant cells, necrosis or granulomas were seen in the sections. Thus a final diagnosis of EG was given.

DIFFERENTIAL DIAGNOSIS
In the context of the present case, osteomyelitis, EG, non-Hodgkin’s lymphoma and Ewing’s sarcoma were considered as clinical differentials. A rare possibility of squamous cell carcinoma of the alveolus was also considered, taking into account the non-healing ulceroproliferative gingival lesion.

The presence of fever, the age of the patient and the gingival ulceration accompanied by bone destruction on radiography favoured osteomyelitis, but the lack of response to antimicrobials and absence of bone necrosis, polymorph infiltration and granulomas on histology ruled out this possibility. Lymphoma was considered due to the presence of fever, pain over the
Eosinophilic granuloma of bone is a rare disease that has a predilection to involve the axial skeleton in children.

In the head and neck region, bony lesions are frequently associated with adjacent soft tissue involvement.

The clinical and radiographic features of eosinophilic granuloma in the jaw are not specific and tend to simulate a host of inflammatory as well as neoplastic conditions, resulting in a diagnostic dilemma.

A high index of suspicion combined with histopathological examination is of the utmost importance in the correct diagnosis of this condition.
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