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Case Report

Rare case report of huge inflammatory pseudotumor of the mandible

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ABSTRACT

Inflammatory pseudotumor (IPT) is a rare benign non-neoplastic lesion. The etiology and the pathogenesis are yet to be understood. Aggressive progression, together with confusing radiological signs, is often mistaken as malignant. We report here a rare case of aggressive IPT in the mandible. In this case a neoplastic lesion or a systemic disease was initially suspected after incisional biopsy. Initial diagnostic treatment with steroids showed a little improvement, which indicated a reactive lesion, IPT. After steroid administration, the residual lesion was resected. Eighteen months postoperative follow-up showed neither recurrence nor progression.

1. Introduction

Inflammatory pseudotumor (IPT), a rare benign non-neoplastic lesion, is composed of a variable numbers of myofibroblastic spindle cells and inflammatory infiltrates of plasma cells, lymphocytes, eosinophils [1]. Although the lung is the most common site of occurrence [2], it has been found to occur in a variety of organs including the major salivary glands and oral cavity [3–6]. The etiology and pathogenesis of IPT, whether it is a result of inflammatory reaction, or truly neoplastic, still remains unclear. Until 1998, such tumors were called as IPT, although Narla et al. [3] later proposed neoplastic variant 'inflammatory myofibroblastic tumor' (IMT) as a more descriptive name. However, it is difficult to distinguish between IPT and IMT. Due to its clinically aggressive behavior along with distractive radiological signs, IPT is often mistaken as malignant. Although a rare condition, progression is rapid and aggressive in the maxillofacial region; hence, early intervention is necessary. Because of its clinical resemblance with malignant legions, it is very important to confirm the diagnosis of such lesions as IPT or IMT for effective management. In this context, immunohistochemically, Coffin et al. [7] demonstrated in 2011 that nearly 36–60% IMTs cytogenetically express anaplastic lymphoma kinase (ALK) protein triggered by ALK gene (at 2p23) rearrangement; therefore, IMT can be characterized as a neoplastic lesion. Here, we present a rare case of aggressive IPT in the mandible.

2. Case report

A 61-year-old male patient presented with swelling, trismus, pain, and feeling of warmth in his left cheek in May 2013. In another hospital he was diagnosed with dentigerous cyst of left lower wisdom tooth and inflammation entrained thereafter, and was treated with antimicrobials. However, his symptoms rather worsened over time, and finally he was referred to the Department of Oral-Maxillofacial Surgery, Dentistry and Orthodontics at Tokyo University Hospital, Tokyo, Japan in August 2013.

Fig. 1A shows diffuse swelling and redness around the surroundings and submandibular lymphadenopathy. Intraoral examination revealed a hemorrhagic soft mass in his left mandibular molar and buccal mucosa region (Fig. 1B). This presented with trismus, only allowing maximum mouth opening (MMO) of only 6 mm. Paresthesia of his lower lip was not clear. Devoid of significant family medical history, he was only present with hypertension and asthma. Hematological investigations showed mild leukocytosis (10,000 cells/μl) and a high serum level of aspartate aminotransferase (AspAT) (formerly known as glutamic-oxaloacetic transaminase or GOT) (51 U/L), alanine aminotransferase (ALT) (also known as glutamic-pyruvic transaminase or GPT) (129 U/L), γ-glutamyltransferase (GGT) (107 U/L), C-reactive protein (CRP:

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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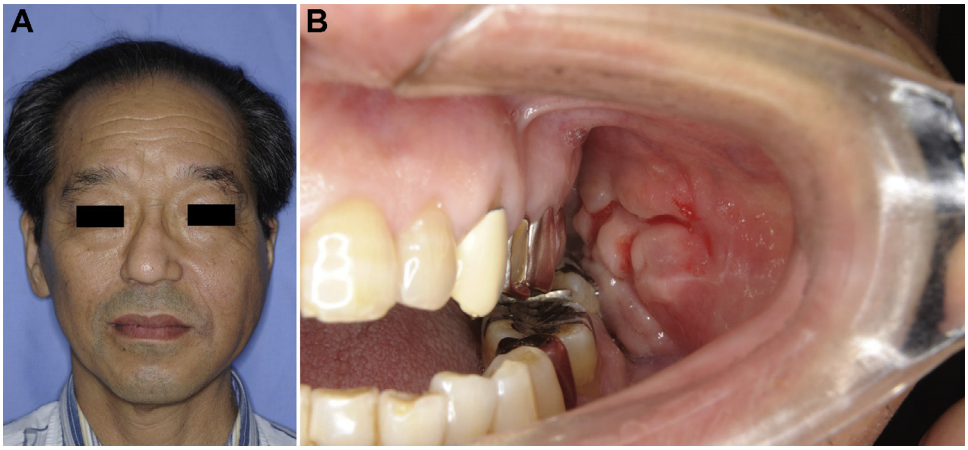

Fig. 1. (A) Frontal view of the face; (B) Image of the oral cavity.

7.65 mg/dL), erythrocyte sedimentation rate (ESR: 118 mm), immunoglobulin G4 (IgG4: 258 mg/dL). Panoramic radiography revealed a radiolucency around the left lower wisdom tooth (Fig. 2A), while computed tomography (CT) scan showed a well-defined osteolytic lesion, extending to the surrounding of left mandibular condyle (Fig. 2B). Gadolinium-enhanced T1-weighted magnetic resonance imaging (MRI) revealed hypertrophy at the left masseter muscle (Fig. 2C and D); the size was 75 mm × 45 mm × 30 mm. Positron emission tomography (PET) with fluorine-18 fluorodeoxyglucose ((18)F-FDG) showed strong accumulation with maximum standardized uptake value (SUV max) of 11.3 around the left mandibular bone and 6.5 around the rectum (Fig. 2E and F). These clinical findings were suggestive of a non-specific inflammatory change or hematological lesion.

Empiric antibiotic therapy (cefcapene, 300 mg/d) for 3 weeks provided no symptomatic improvement. Then we performed an intraoral incisional biopsy of the mandibular lesion in September 2013. Histopathological analysis revealed a broad and dense hyperplastic lesion of collagenous fibers, accompanied by inflammatory cell infiltration, mainly lymphocytes and plasma cells. The inflammatory fibrotic lesion infiltrated the striated muscle undergoing degeneration. The growing fibroblast-like cells were atypical, and mitosis was not prominent (Fig. 3). These characteristics were consistent with IPT, although it was difficult to differentiate from a reactive lesion, a neoplastic lesion, or a generalized disease, such as IgG4-related sclerosing disease (IgG4-RD). Next, corticosteroid (prednisolone) at 30 mg/day (0.6 mg/kg/day) was administered *per os* (PO) for 2 weeks to provide diagnostic treatment. In the posttreatment evaluative (18)F-FDG PET/CT, the SUV max of the mandibular lesion was reduced to 6.5 and the initial rectal accumulation disappeared. Serum level of IgG4 also decreased to 210 mg/dL. Clinically, there was improvement of the swelling and trismus; MMO became 12 mm. Corticosteroid administration showed some efficacy in anti-inflammation action, though CT and MRI scans failed to show improvement in mass (Fig. 4). Corticosteroid administration was gradually weaned over a week. The poor response to corticosteroids indicated it was much more likely to be reactive rather than inflammatory, and we decided on its resection.

The resection with the disarticulation of the mandible was performed posterior to the left second molar including the mandibular condyle under general anesthesia in January 2014. The tumor was yellowish-white with rubbery fibrous scar tissue, infiltrating and adhering strongly to surrounding normal soft tissue (Fig. 5A and B). The histological examination showed a fibrotic lesion with a wide infiltration of plasma cells and lymphocytes spread throughout intraosseously and extraosseously (Fig. 6A). Immunohistochemical investigation revealed focally positive spindle cells for α-smooth muscle actin and caldesmon while negative for desmin, anaplastic lymphoma kinase and ALK (Fig. 6B and C). The ratio of IgG4+/IgG+ plasma cells was less than 10%. From these morphologic and immunohistochemical findings, we finally diagnosed the lesion as IPT. Postoperatively, trismus improved prominently; MMO became 32 mm. (18)F-FDG PET examination a year postoperation showed a SUV max of 2.1 at the left masseter, which indicates physiological muscle activity and disappearance of abnormal accumulation (Fig. 7A and B). Serum level of IgG4 decreased to 113 mg/dL 2 months postoperation (May 2014), and entered within normal range of 80.5 mg/dL a year after (January 2015). There has been no radiological or clinical proof of any recurrence or progression of the disease up to July 2015.

3. Discussion

The variations in the histological appearance suggest that the term IPT does not represent a single entity, but a group of lesions demonstrating nonspecific chronic inflammatory changes [9]. IPT is prevalent in the lung, whereas head and neck occurrence accounts for only 5% of IPT, mostly in the buccal mucosa, mandible and tongue [10,11]. The etiology and pathogenesis of IPT are yet to be fully explained; however, after complete resection of such lesions, systemic symptoms characteristically seen in IPT, such as pyrexia, decreased body mass, local pain, anemia, thrombocytosis, leukocytosis, are improved [12]. Although the mechanisms involved in developing these symptoms are poorly understood, overproduction of interleukin (IL)-6, IL-8, IL-1β, and monocyte chemotactic protein-1 might induce inflammatory cell migration and accumulation [13–15]. In other hypotheses, the immunological host reaction to stimulants such as microorganisms, adjacent necrotic tissue, neoplasm, foreign bodies, some type of tissue injury [16], Epstein–Barr virus infection [17], and chronic irritation come to cause the legion. In our case, the resected specimen showed no cystic lesion and chronic pericoronitis of wisdom tooth may be associated with oncogenesis.

These reactive lesions account for a substantial portion of IPT, and truly neoplastic counterpart of IPT is called IMT [18,19]. It arises mostly in lung, mesentery, omentum, and has a benign course. It has intermediate biologic potential and the recurrence rate varies at different sites from 2 to 25%, and develops distant metastasis in less than 5% [20]. On the other hand, IPT confined to a single