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A case of primary osteomyelitis of the mandible preceding Takayasu arteritis

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ABSTRACT

Primary chronic osteomyelitis (PCO) is a rare condition that usually affects the mandible. Although most often occurring isolated, it can be part of various systemic and cutaneous syndromes. Takayasu arteritis (TAK) is an autoimmune disease that can co-exist with osteomyelitis, but has only been reported once in conjunction with PCO of the mandible. We present a case of a 27-year old female with PCO of the mandible which was found to have co-existing TAK. Doctors treating patients with PCO should be aware of the associated diseases and actively seek out their signs and symptoms.

1. Introduction

Primary chronic osteomyelitis (PCO) of the mandible is a rare condition.

We present a 27-year old pregnant woman with PCO of the mandible found to have Takayasu arteritis (TAK). To this date, there are seven cases reported in the literature of patients suffering from osteomyelitis of non-craniofacial bones that also developed TAK, but only one patient with osteomyelitis of the mandible [1]. Additionally, there is only one case report of a patient with osteomyelitis of the mandible during pregnancy [2].

This case report will give the reader an overview of PCO of the mandible and the different syndromes and disease association.

2. Case report

2.1. Initial presentation and treatment at the maxillofacial surgery department

A 27-year old woman was referred to the Maxillofacial Surgery Department at Oslo University Hospital, Ullevål. The patient had no past medical history. She was 15 weeks pregnant, expecting her first child. The third molar in the left mandible had been surgically extracted four years prior because of pericoronitis after which there was a brief period of surgical wound dehiscence. The third molar in the left maxilla was extracted 11 months before presenting to the department. She reported having had diffuse pain in the left jaw area four months ahead of the removal of the tooth. The pain had gradually moved to the left hemi-mandible which had increased in size the

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last couple of months. Panoramic radiograph (Fig. 1) and cone beam computer tomography done at her dentist showed cortical thickening and signs suggesting long-standing inflammation in the left hemi-mandible compatible with osteomyelitis.

On examination, the patient had a bony swelling at the body of the left hemi-mandible from the first pre-molar to the second molar. The swelling was tender to palpation, but there were no other obvious signs of infection such as redness, fistula or increased skin temperature. Thus, antibiotic treatment was not initiated. A left-sided submandibular lymph node was tender to palpation and slightly increased in size. She had hypoesthesia on the chin and lip on the left side as a sign of involvement of the inferior alveolar nerve. There were no findings on the intraoral examination.

The patient was closely followed up during her pregnancy, and as there was no clinical detriment, we decided to wait until after birth before considering surgical treatment. After uncomplicated birth, the patient underwent a CT scan which showed what appeared as a chronic diffuse sclerosing osteomyelitis from the first premolar to the ramus of the left hemi-mandible (Fig. 2). It was decided to do a decortication because of the substantial swelling on the lower left side of the face.

The patient was admitted for operation 11 months after her first visit to the Department. On pre-operative examination, a bruit was noted on her left carotid artery. Laboratory samples showed anemia with hemoglobin 10.6 g/dL and increased inflammatory parameters with ESR of 73 mm/h [1–17], C-reactive protein of 23 mg/L (<4), and leucocytes at 13 cells per mcL (3.5–10). An ultrasound showed slightly increased speed (100–110 cm/sec) in the common carotid arteries (CCA) and internal carotid arteries (ICA) on both sides. The left vertebral artery appeared smaller in diameter than the opposite side, but the speed was equal (65 cm/sec). CT angiography showed increased tissue density in the aortic arch wall, a narrow left CCA, and high-grade stenosis of the left subclavian artery (Fig. 3). The stenosis of the subclavian artery continued to the left vertebral artery, that was narrowed in its whole length. A tentative radiological diagnosis was vasculitis of the aortic arch and major precerebral arteries most compatible with TAK.

The patient was operated the following day with buccal decortication from the first premolar to the ascending ramus of the left hemi-mandible. Intraoperatively, there were scarce amounts of trabecular bone and the bone marrow appeared less vascularized than normal. Histology showed chronic inflammation with massive apposition of cortical bone and scarce amounts of trabecular bone in which there was noted fibrosis and few plasma cells. There were no adverse complications.

The patient was transferred for follow-up of the vasculitis at the Department of Rheumatology.

2.2. Follow-up and treatment at the department of rheumatology

The patient had experienced unspecific symptoms of fatigue, general weakness, increased sweating, and increasing numbness in the left hand for three years. There were no definite symptoms of ischemia in the left arm. Her left-sided facial pain was regarded as atypical of carotidynia, but she had over time developed jaw claudication that improved during the pregnancy. Inflammatory markers were increased with ESR 107 mm/h [1–17], and C-reactive protein at 57 mg/L (<4). She was anemic with hemoglobin 9.8 g/dL (11.7–15.3), interpreted as a combination of iron deficiency and anemia of chronic disease. On examination, there was a bruit over the left carotid arteries and a blood pressure difference of more than 20 mmHg as well as reduced pulse amplitude in the left radial artery.

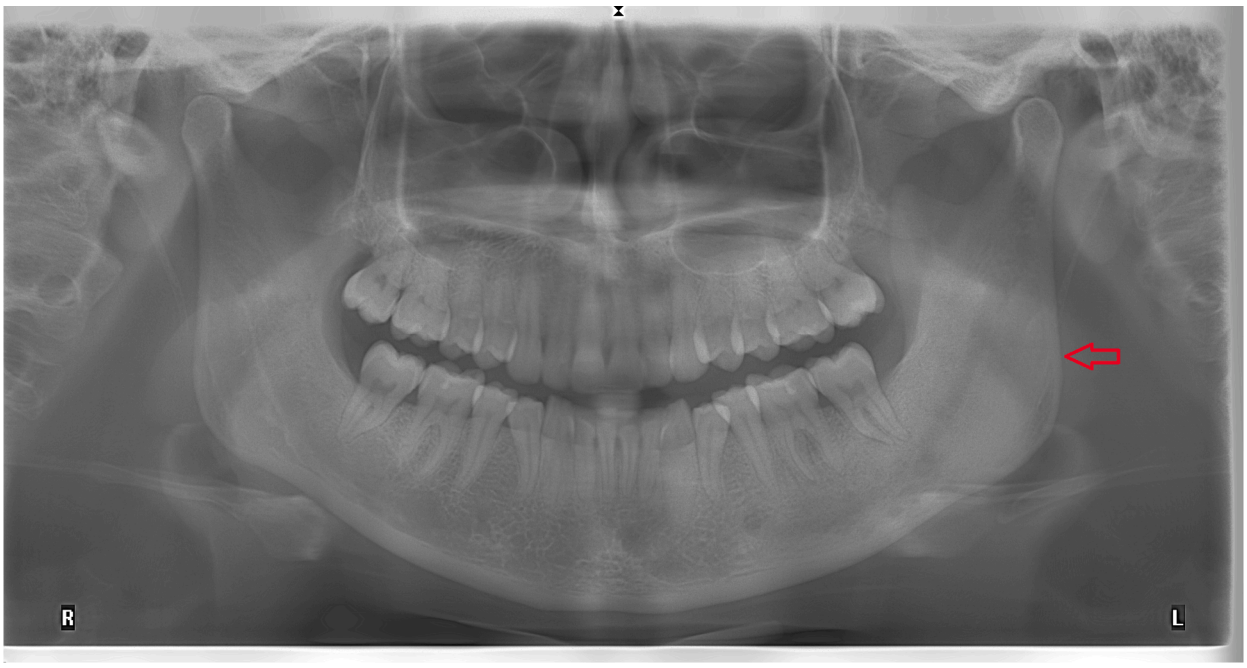

Fig. 1. The panoramic radiograph shows subtle osteosclerosis and slight cortical thickening (red arrow) in the left hemi-mandible, extending from the body to the ramus.

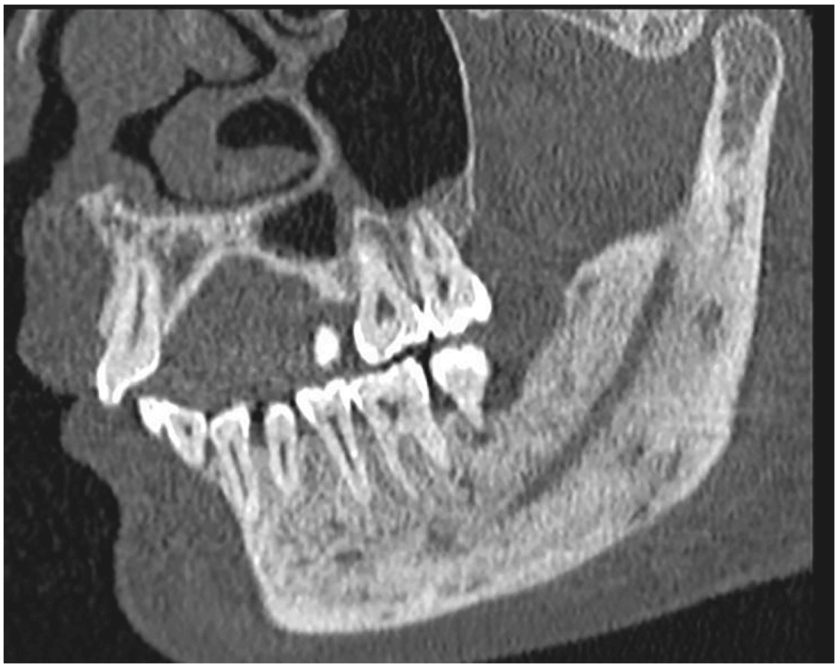

Fig. 2. CT of the mandible showing homogenous sclerotic bone of the left hemi-mandible, including the mandibular ramus, while the condylar process appears normal.

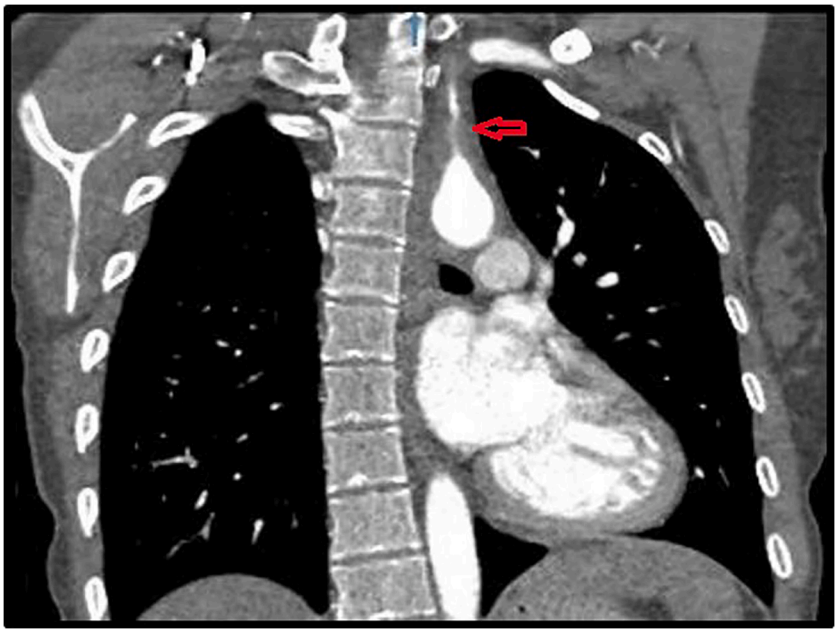

Fig. 3. CT arteriography showing distinct luminal narrowing and thickened arterial wall of the left subclavian artery proximally (red arrow).

CT angiography was repeated three days after the initial one, which showed wall thickening of the aortic arch, the thoracic artery, and precerebral arteries. There was high-grade stenosis of the left subclavian and left vertebral artery and wall thickening of the left CCA. A final diagnosis of type IIb Takayasu arteritis was made, with the involvement of the descending thoracic aorta, the aortic arch and its branches [3].

Treatment was initiated with intravenous methylprednisolone 750 mg once daily over three days followed by oral prednisolone 1 mg/kg daily and subcutaneous methotrexate 20 mg once weekly. The treatment resulted in rapid recovery of her symptoms and normalization of the inflammatory parameters. Three months later, prednisolone had been tapered down to 12.5 mg/daily. An 18F-

labeled fluoro-2-deoxyglucose (^{18}F-FDG) positron emission tomography showed slight FDG in the vessel walls of the aortic arch, the brachiocephalic trunk, the left proximal common carotid, and the left subclavian artery (Fig. 4). There were no signs of inflammation of the mandible. The radiological results corresponded with the finding of a consistent difference in the blood pressure of about 20 mmHg between the left and right upper arm.

Further tapering of oral prednisolone is scheduled and follow-up at the Department of Rheumatology. The left hemi-mandible remains asymptomatic and there are no signs of facial asymmetry.

3. Discussion

There is a debate about the definition, etiology, and classification of the different types of osteomyelitis of the mandible [4]. Eyrich et al. introduced the Zurich classification system, separating the disease in an acute and chronic form based on the duration of less- or more than one month. They further made a distinction between a secondary chronic type which succeeds the acute form, and a primary chronic type which is inflammatory and non-suppurative [5]. The latter is often used synonymously with diffuse sclerosing osteomyelitis which again has been denominated under various names [4]. The disease often has an insidious occurrence without an acute phase and is characterized by intermittent pain, swelling, lymphadenopathy, and hypoesthesia in the area provided by the inferior alveolar nerve, all of which our patient had to some extent [6]. Also, patients may have trismus, but our patient did not experience this [6]. PCO does not have the formation of pus, fistula or sequestration as opposed to chronic secondary osteomyelitis and osteoradionecrosis [7]. The etiology of PCO is unclear, although there have been multiple different suggestions such as a subacute infection, craniofacial muscle overuse, and autoimmune mechanisms [4,8]. Radiographic findings include osteolysis, osteosclerosis, subperiosteal bone formation, and occasionally hypertrophy of the masseter muscles [7]. Histological features may in various degrees include subperiosteal formation of new bone, bone resorption, increased volume of trabecular bone, medullary fibrosis, microabscesses, lymphocytes, plasma cells, and hyalinosis around small vessels [5,9]. Treatment options are decortication, antibiotics, corticosteroids, NSAIDs, hyperbaric oxygen treatment, bisphosphonates, and RANKL inhibitors [5,10–12]. Decortication involves removing ill-perfused, dense cortical bone in order to ameliorate blood-supply and thereby accelerate healing [13–15]. In our patient, decortication was also performed with the aim of decreasing the volume of the mandible.

PCO most commonly occurs isolated but is also associated with synovitis, acne, pustulosis, hyperostosis and osteitis syndrome (SAPHO), and chronic recurrent multifocal osteomyelitis (CRMO) [16,17]. The latter most often occurring in children and the former in adults. Furthermore, there is an association between CRMO and various dermatological conditions such as pustulosis palmoplantaris, psoriasis vulgaris, acne, generalized pustulosis, pyoderma gangrenosum, and non-cutaneous inflammatory conditions such as inflammatory bowel disease, celiac disease, inflammatory arthritis, dyserythropoietic anaemia, sclerosing cholangitis, parenchymal lung disease, sacroiliac joint involvement, Still's disease, Ollier disease, Sweet syndrome, and ANCA-positive vasculitis [18].

TAK is a large-vessel vasculitis, and it primarily affects the aorta and its primary branches. TAK shares histopathology and clinical features with giant cell arteritis (GCA, also known as temporal arteritis), the other large-vessel vasculitis [19]. Distinction between the two disorders can usually be made based on patient age and distribution of lesions. GCA affects patients 50 years and older, and TAK occurs in younger patients with a female:male ratio of about 9:1. The pathogenesis is poorly understood. The onset of TAK tends to be subacute, ranging from months to years. Our patients long-standing symptoms in the left arm, the jaw claudication and systemic symptoms indicate that the disease probably was present years prior to diagnosis. It is thus likely that the debut of TAK preceded PCO.

A recent epidemiological study from Norway showed that a point prevalence of Takayasu arteritis segregated by ethnic origin was 22.0 per 10^6 (95% CI; 17–29) in those of native Scandinavian ancestry. The prevalence was higher in those of Asian, 78 per 10^6 (95% CI 38–152) and of African descent, 108 per 10^6 (95% CI 46–254). The recent incidence rate in Norway is estimated to be two per million per year [20].

TAK is associated with inflammatory bowel disease (IBD) and ankylosing spondylitis. In the Norwegian cohort, IBD was observed in 8% and AS in 7%, both far exceed what would be expected by chance. Also, there are several case reports of the association of pyoderma gangrenosum and erythema nodosum and TAK [21].

It is not likely that the bone involvement in our patient was directly caused by her vessel involvement and ischemia. The most likely cause is a common pathogenic immunopathology. The symptoms of jaw claudication and left-sided hand numbness was probably caused by ischemia as a result of luminal narrowing of the ipsilateral common carotid and subclavian artery.

A recently published case report by Shirai et al. found only six published cases of chronic osteomyelitis occurring with TAK and reported a prevalence of osteomyelitis in adult patients with TAK of 1.47% [1]. Further presenting two additional cases in their report, the total amount of published cases, including our patient, making nine in total. Eight of these nine patients were women with a mean age of 25,5 years. Only one of the eight previously published cases had chronic osteomyelitis of the mandible in occurrence with TAK, while the rest had involvement of other bones [22–27]. Additionally, there is only one earlier report of osteomyelitis of the mandible during pregnancy [2]. Our case shares several findings with the case presented by Shirai and colleagues. Both patients were females in their twenties, had symptoms in the lower jaw and neck, and both responded well to treatment with prednisolone. Our patient did, however, have a history of third molar extraction following pericoronitis, which was not present in the case report by Shirai et al. It is possible that the extraction may have introduced bacteria in the mandible which subsequently could have initiated the inflammatory changes. This is, however, unlikely as there were no signs of infection other than pain and swelling, which can be explained by sterile inflammation. The inflammatory changes in the mandible were likely caused by the same immunological process that caused the vasculitis, rather than being a result of hypoxia or infection [1].

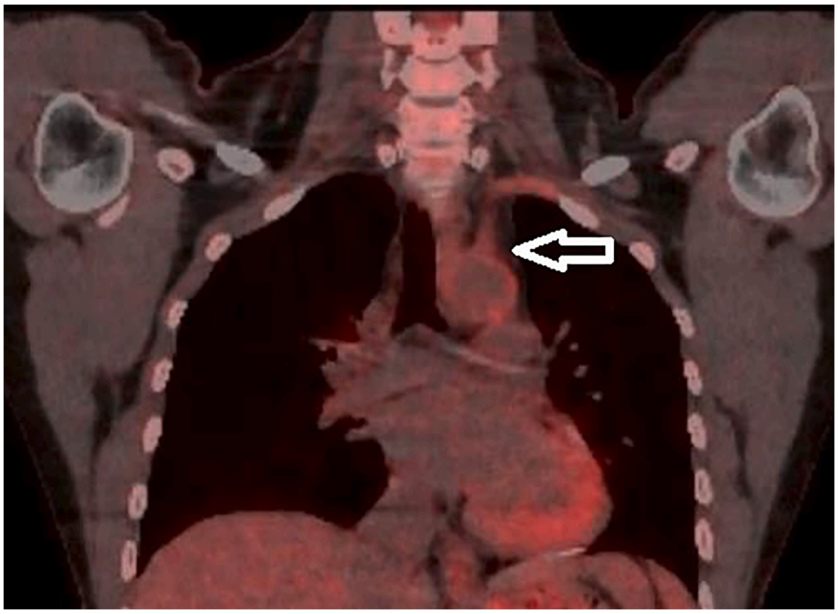

Fig. 4. PET showing increased uptake in the wall of the aortic arch and proximal left subclavian artery (white arrow).

4. Conclusion

We present the second case of a patient with primary chronic osteomyelitis of the mandible associated with TAK. Our case report illustrates the importance of awareness of the possible coexistence of other disease entities in patients suffering from chronic osteomyelitis of the mandible.

Conflicts of interest

None.

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