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CASE REPORT

Long-Term Follow-Up of Bimaxillary Osteomyelitis Associated with Autosomal Dominant Osteopetrosis: A Case Report

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Abstract Osteopetrosis (OP) is a rare metabolic bone disease characterized by a generalized increase in skeletal mass. The disease is characterized by increased susceptibility to develop osteomyelitis of the jaws. We report a case of clinical and radiological progression of bimaxillary osteomyelitis in a patient with autosomal dominant OP. The patient presented non simultaneous osteomyelitis in both upper and lower jaws with time interval of 10 years. The osteomyelitis of maxilla resulted in oroantral fistula formation and required surgical closure with Bichat fat pad flap. The mandibular osteomyelitis resulted in sequestra formation and pathological fracture and required multiple debridement procedures. Both maxillary and mandibular osteomyelitic foci were persistent and healing required more than 2 years. The case confirms the possibility of development of non simultaneous bimaxillary osteomyelitis in patients with OP. The case enhances the need of close follow- up and preventive measures in patients with OP.

Keywords Osteopetrosis · Osteomyelitis · Jaw bones

Introduction

Osteopetroses (OP) are generalized bone dysplasias caused by a congenital defect in the development or function of the osteoclasts resulting in generalized increase in skeletal mass with reduction of marrow spaces (bone).

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Clinical classification is difficult due to the high variability of severity and associated complications. Three main phenotypes of OP are described in the literature: an infantile malignant autosomal recessive form, an intermediate autosomal recessive form, and an adult benign autosomal dominant form [1, 2]. Autosomal dominant osteopetrosis (ADO) is the most common type of OP with estimated incidence of 5 in 100,000 births. ADO type 2 is related to chloride channel 7 gene mutation CLCN 7 which encodes a chloride channel found only in osteoclasts [3, 4].

ADO type II is the most common form and has an extremely heterogeneous course ranging from an asymptomatic to a severe phenotype. The main clinical findings of ADO are fractures of long bones and osteomyelitis of the mandible. Bone involved with OP is thought to have a compromised vascular supply and therefore to be more susceptible to osteomyelitis.

The present article reports a case of bimaxillary non simultaneous osteomyelitis associated with ADO. The clinical and radiological patterns and progression over long term follow-up are discussed.

Case Report

The patient, 66 years old male, was referred to our department for evaluation of an oroantral fistula which resulted from the third upper right molar extraction. The extraction was performed by dental practitioner 2 weeks before the consultation.

Patient history revealed that he had a previous diagnosis of OP in adulthood. He suffered bony fractures of humerus and femur (Fig. 1) and also presented hearing impairment and myelophthisic anemia.

Fig. 1 Transverse fracture of the right femoral bone

Intraoral examination revealed the alveolar socket created after removal of the third upper molar with no signs of healing. The area of bone exposure was <5 mm and the oroantral fistula was present. The patient was primarily treated conservatively with administration of prolonged courses of antibiotics (1 month) and topical decongestives. This treatment was not effective and the area of bone exposure was increasing. The diagnosis of maxillary osteomyelitis associated with autosomal dominant OP was done. Five months later the patient did not present any signs of healing and signs of chronic sinusitis were observed. The surgical treatment for closure of the oroantral fistula and area of bone exposure was undertaken. The fistula was closed with pediculated flap of Bichat's buccal fat pad under general anaesthesia. The patient presented no signs of oroantral communication neither bone exposure in the early postoperative period but 3 months later recurrence occurred. The larger area of bone exposure in the right maxillary alveolar ridge was observed with pathologic discharge and severe local pain. The facial CT scan revealed large area of bone destruction in the right maxilla and sequestrum formation (Figs. 2, 3). CT scan also revealed typical signs of OP as generalized sclerosis of facial bones, sinus obliteration and endobones formation (Fig. 4). The sequestrum was removed under general anaesthesia. The pathology report confirmed the osteopetrotic changes in the removed bone, the diagnosis of ADO was confirmed. The patient was followed up and received various courses of antibiotics and curettages. The complete closure of the bone exposure in the right maxilla was observed only 3 years after the tooth extraction.

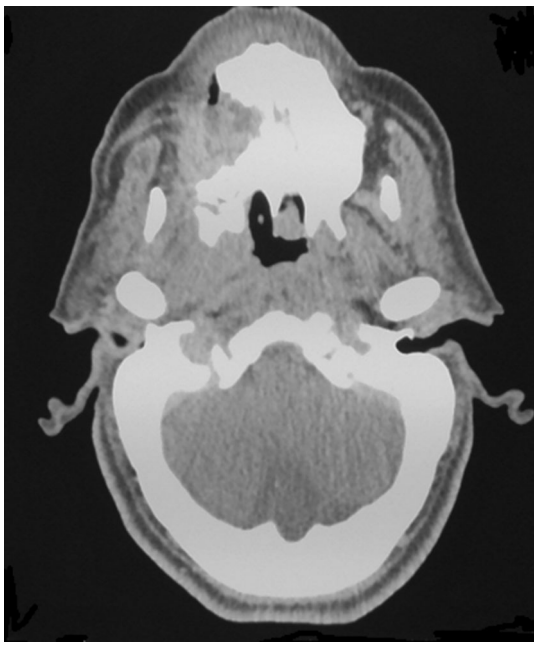

Fig. 2 CT scan showing the large area of bone destruction in the right maxillary bone

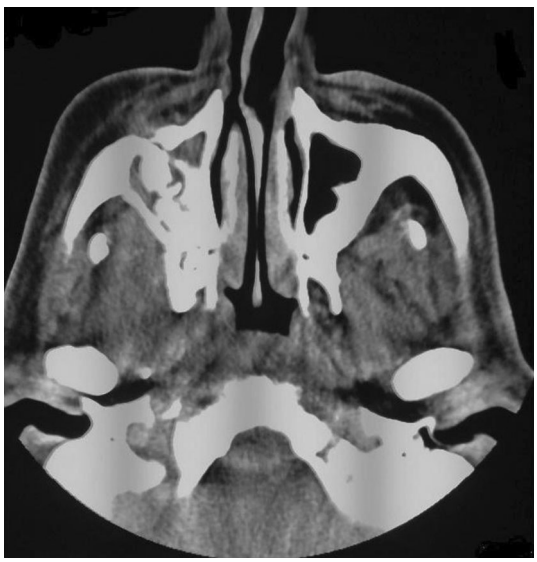

Fig. 3 CT scan showing formation of sequestrum in the right maxillary bone

Seven years later the patient came back to our department with a new osteomyelitic focus in the left mandible after left inferior mandibular molars extraction. The panoramic radiogram revealed signs of osteomyelitis of the lower jaw bone (Fig. 5). The patient received curettage treatment and various courses of antibiotics without significant improvement. Two years later sequestrum formation in the zone of bone exposure was observed on the panoramic radiography (Fig. 6). CT scan confirmed sequestrum formation,

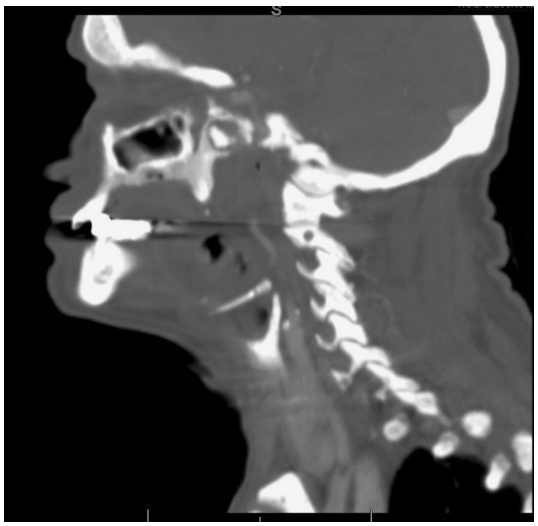

Fig. 4 CT showing endobone formation in the sphenoid bone

pathologic fracture of the mandible and inflammatory changes in masticator space (Fig. 7). Two months later the patient was admitted to emergency department with symptoms of important left hemifacial swelling and trismus (Fig. 8). The urgent contrast enhanced CT revealed formation of an abscess in the left masticator space. The abscess was surgically drained via skin incision, a large amount of purulent discharge was obtained and a rubber drain was placed in the site of incision. The patient required in patient stay and intravenous antibiotic treatment. Rapid clinical improvement was observed and the patient was discharged. In the follow-up a cutaneous fistula formation in the site of surgical incision was observed (Fig. 9). The complete healing of osteomyelitic focus with closure of the bone exposure in the left mandible and closure of the chronic fistula was observed 2 years after teeth extraction. The patient lost significant height of the alveolar ridge in the left mandible bone. Final panoramic radiography revealed bone mass loss in the left mandible and the right maxilla (Fig. 10).

Discussion

The patients with ADO have increased susceptibility to develop osteomyelitis of the jaws. Ten percent of OP cases

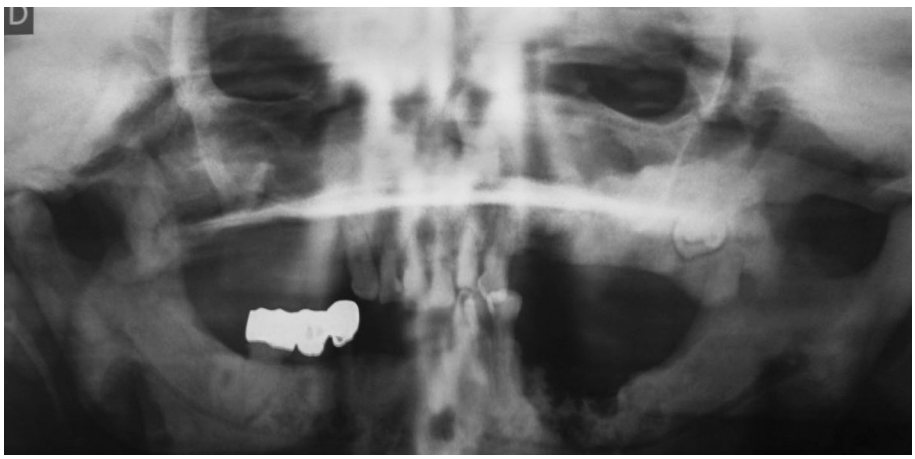

Fig. 5 The panoramic radiogram shows signs of osteomyelitis of the lower jaw bone

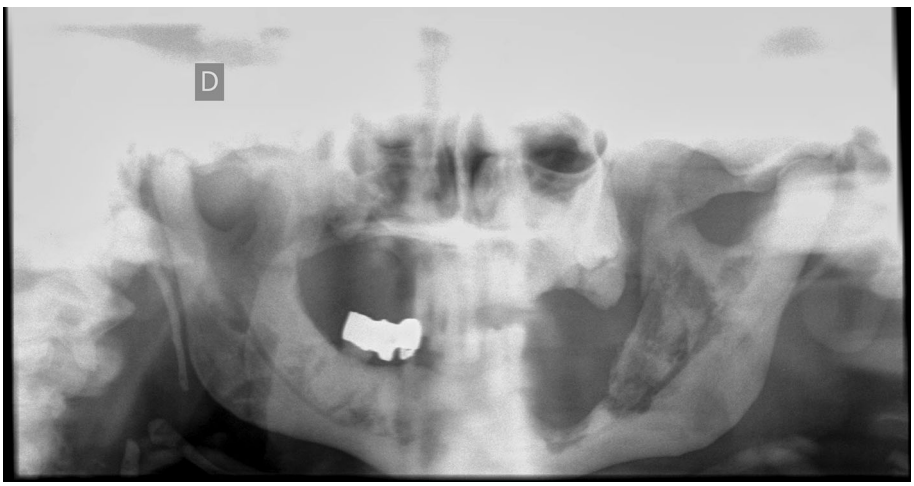

Fig. 6 The panoramic radiogram shows sequestrum formation in the left lower jaw bone

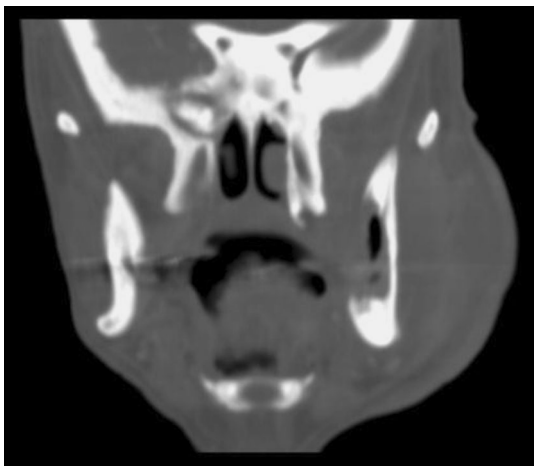

Fig. 7 CT scan showing pathological mandibular fracture

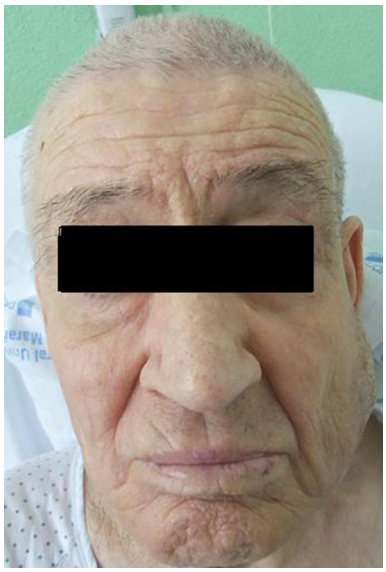

Fig. 8 Abscess in the left masticator space: important hemifacial swelling

develop osteomyelitis that generally involves the mandible [5]. Several cases of simultaneous bimaxillary osteomyelitis have been reported [6]. We report the case of non simultaneous bimaxillary osteomyelitis in a patient with ADO developed with time interval of 10 years. The clinical and radiological progression of the disease over 12 year interval is presented. To our best knowledge this is the longest follow up observation of the patient with maxillomandibular osteomyelitis associated with ADO reported in the literature.

The ADO is a heterogenous genetic disorder that can present the following signs and symptoms: bone sclerosis, cranial nerve compression with blindness and deafness, recurrent long bone fractures, myelophthisic anemia. In head and neck the classical manifestations are: frontal

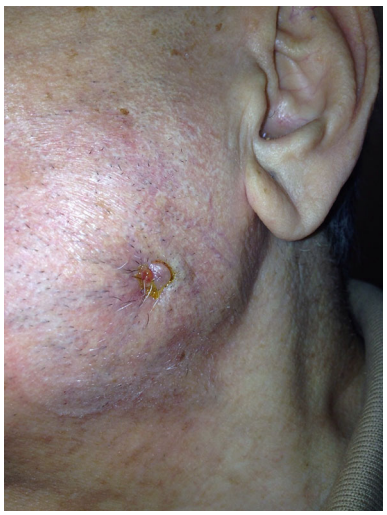

Fig. 9 Cuataneous fistula formation after drainage of the abscess in the left masticatory space

bossing of the skull, delayed teeth eruption, early loss of teeth, congenitally missing or malformed teeth and thickened lamina dura and increased susceptibility to develop osteomyelitis of the jaws [7].

In our patient we observed typical signs of OP such as long bone fractures, sensorineural hearing loss, myelophthisic anemia and bimaxillary osteomyelitis.

The differential diagnosis of OP should be done with Paget's disease, pycnodysostosis, polyostotic fibrous dysplasia, osteomyelitis occurring in a gigantiform cementoma, fluorosis, generalized carcinomatosis of bone, and osteomyelosclerosis, craniometaphyseal dysplasia, diaphyseal dysplasia (Camurati Engelmann–Ribbing disease), melorheostosis, and osteopathia striata [8].

Bone biopsy is not essential for diagnosis because radiological findings and history of frequent long bones fractures are usually diagnostic [6].

The main radiological finding in ADO is general increase in skeletal radiodensity. Thickening of vertebral endplates with sandwich vertebra pattern is typical. Bone within bone pattern has been reported in 94.4 % of patients [9, 10]. The 'bone within bone' or endobone finding differentiates OP from other sclerosing dysplasias and it is due to the cyclical nature of the disease, so that the dense shadow of bone at the time of formation of abnormal bone is seen within the outline of the current normal or abnormal shadow [8]. Endobone formation is most commonly observed in the iliac wings.

In head and neck region the radiological signs of OP are: increased density of the bones in the skull base, increased density of orbital margins, sinus obliteration.

The endobone formation is a rare finding in the head and neck region. In our case formation of endobone was observed in the skull base in the wings and body of sphenoid

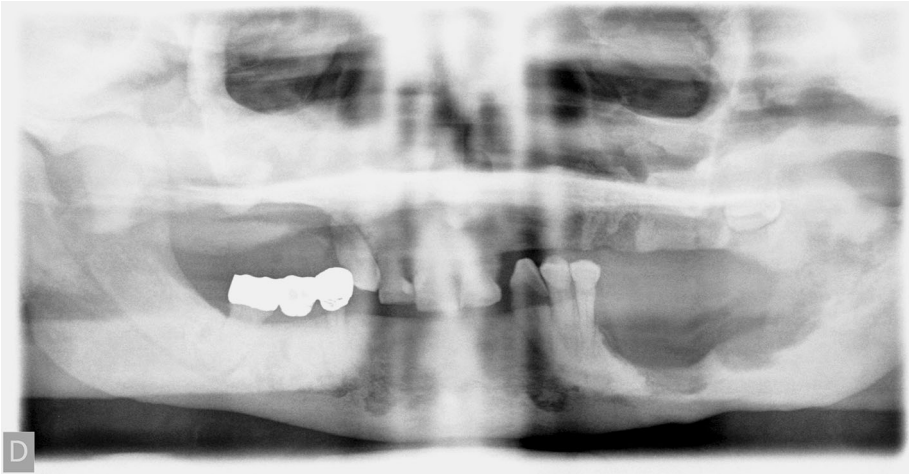

Fig. 10 Final panoramic radiograph. *D* denotes right side

bone. The dental abnormalities, such as delayed teeth eruption, hypodontia, hypercementosis and odontomas can be revealed on panoramic radiography.

The signs of maxillomandibular osteomyelitis are supportive for diagnosis of ADO: in maxilla trabecular bone defect can be observed, in mandible osteosclerosis, periosteal reaction, cortical bone defect and sequestra formation are common findings [11]. In cases of osteomyelitis pathological fracture of the mandible can occur and can be observed on radiological examination. The radiological evolution of osteomyelitis in our case shows progressive bone loss due to sequestra formation and discharge and multiple debridement procedures (see Fig. 5, 10).

Osteomyelitis is an inflammatory condition of bone involving both medullary bone, cortex and haversian system. It has been outlined that in cases of osteomyelitis predisposing bone pathology and immunosuppressive conditions should be ruled out [12]. In our patient both the pancytopenic condition and OP contributed to development of the osteomyelitis.

The underlying deficient bone vascular supply and immunosuppressive condition explains the torpid course of osteomyelitis in patients with OP. In our patient the osteomyelitis affected primarily the upper jaw and in 10 years the lower jaw. In both cases healing took around 3 years and in both cases it was caused by teeth extractions.

Surgical options of treatment of osteomyelitis in OP patients include marginal mandibular resection, closure of oroantral fistula and local debridement procedures [5]. Fibular flap mandibular reconstruction has been reported in a patient with OP [13]. Hyperbaric oxygen therapy was advocated as a conservative method of treatment of OP osteomyelitis. In our patient multiple debridement procedures, facial abscess drainage and antimicrobial treatment were carried out for mandibular osteomyelitis. No major surgical treatment was considered due to the general condition of the patient. Maxillary osteomyelitis resulted in oroantral communication formation closed surgically with Bichat fat pad flap.

Caution should be taken in even minor oral surgical interventions in patients suffering from OP and antibiotic coverage can be recommended. Endodontic treatment is preferable to extractions and oral hygiene should be extreme [14, 15].

Conclusions

The clinical and radiological progression of osteomyelitis of the jaws in patient with ADO is described. The radiological signs of OP are very characteristic and have diagnostic value. The case confirms that the OP patients are very susceptible to develop osteomyelitis of the jaw bones, especially as a result of tooth extractions. The osteomyelitis in patients with OP is very persistent and difficult to treat. No effective methods have been described for treatment of jaw osteomyelitis associated with OP. We consider that all the patients with osteomyelitis require close longlife follow up and preventive measures in order to avoid new osteomyelitic foci formation.

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