Recurrent osteomyelitis of the mandible in osteopetrosis: a common complication of an uncommon disease

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DESCRIPTION

Osteopetrosis is a congenital disease characterised by overtly dense bone with obliteration of marrow spaces, owing to defective osteoclast function, resulting in excessive and defective bone formation. It is rare, having an incidence of 1 in 250 000 births.1 Diffuse generalised osteosclerosis of bones is seen and serious oral complications such as osteomyelitis with exposed necrotic bone may occur in the jaws.2 Ten per cent of osteopetrosis cases develop osteomyelitis that usually involves the mandible. Osteomyelitis of the maxilla is very rare, probably because of the thin cortical bone and rich collateral blood supply. Osteopetrotic bones lack a bone marrow cavity, fail to resist normal physical loads, and are vulnerable for fractures due to lack of remodelling.1 Osteopetrosis is generally diagnosed through skeletal radiographs in which there is increased radiodensity and hence the name ‘Marble bone disease’.3 Dentists should be aware of the disease because tooth extraction in such patients results in impaired wound healing with recurrent, recalcitrant osteomyelitis.

A 40-year-old man reported with an 8-month history of pain and pus discharge from the right side of his face. History revealed that he had undergone extraction of carious mandibular molar teeth 3 years prior, following which he had recurrent pus discharge, pain and swelling. Since the extraction site did not heal and the patient experienced pain, he subsequently underwent extraction of all his mandibular teeth within 2 years. Even after extraction of all mandibular teeth there was pus discharge and mandibular fracture. The patient had hip bone fracture 6 years earlier. There was no history of drug use or any other disease, and his personal and family histories were unremarkable.

A diffuse, firm and tender swelling was seen on the right body of the mandible, with two pus discharging sinuses. Intraorally, a single diffuse swelling was seen on the right mandibular alveolus obliterating the buccal sulcus, firm in consistency and slightly tender with discharge of pus on palpation (figure 1). A panoramic radiograph revealed diffuse osteosclerosis of the maxilla with indistinct trabeculae and marrow spaces. A ‘moth eaten’ appearance of the right ramus and angle of the mandible was evident with evidence of a fracture malunion (figure 2).
Figure 3  Posteroanterior (A) and lateral (B) skull radiographs, chest radiograph (C), and radiographs of the upper (D) and lower (E) extremities showing generalised increased radiodensity and Erlenmeyer flask deformity of the tibia (E).

Figure 4  Bony sequestra obtained following curettage. (A) H&E section showing lamellar bone with numerous lacunae devoid of osteocytes and lack of osteoblastic rimming (original magnification ×10); (B) the bony trabaculae also exhibit haematoxyphilic lines with evidence of scanty stroma and areas of vascularity and haemorrhage (original magnification ×40; C and D).
observed (figure 2). Radiographs of the skull, chest and limbs also showed diffuse osteosclerosis with an Erlenmeyer flask deformity seen in the tibia (figure 3).

Pycnodysostosis, craniometaphyseal dysplasia (Pyle’s disease), diaphyseal dysplasia (Camurati-Engelmann-Ribbing disease), osteosclerosis of fluoride poisoning, melorheostosis and osteopathia striata may be considered in the differential diagnosis.

Complete blood count revealed anaemia, neutropaenia and elevated serum acid phosphatase levels. Debridement of the affected site was carried out and bony sequestra were removed (figure 4A). The patient was given clindamycin 300 mg tablets two times per day for a week and kept under regular follow-up every 6 months.

Histopathological evaluation revealed lamellar bone with numerous lacunae devoid of osteocytes and osteoblastic rimming and scanty stroma with areas of vascularity (figure 4A–C). A diagnosis of recurrent osteomyelitis in benign adult osteopetrosis was made and the patient was kept on regular follow-up.

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