Clinical considerations for prosthodontic rehabilitation of intermediate form of osteopetrosis: A report of two cases

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ABSTRACT

Introduction: Osteopetrosis is a rare autosomal hereditary disorder characterized by abnormal functioning of osteoclasts, which leads to delayed wound healing. Hence chances of infection are increased even after minor surgical procedures. This paper aims to describe clinical features, and prosthodontic management of two patients who presented with clinical features of intermediate form of osteopetrosis and missing teeth.

Case report: A 27-year-old patient having intermediate osteopetrosis presented with difficulty in chewing food and swallowing, due to edentulous maxillary and mandibular arches with large antral cavities, which developed secondary to tooth extraction. Another case discussed is a 25-year-old female patient who reported with chief complaints of difficulty in chewing due to resected left side of lower jaw. In these case reports, importance of avoiding extractions, preservation of remaining teeth and conservative management with minimum number of patients' visits has been emphasized.

Conclusion: Efforts should be made to avoid dental extraction in such cases as chances of bone infection and fracture are more. If patient is partially or completely edentulous only removable type of prosthesis is advisable as bones are hypocalcified and maintenance of oral hygiene is easy with removable prosthesis.

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INTRODUCTION

Osteopetrosis was first described in 1904 by Albert Schonberg.¹ Osteopetrosis, also known as Marble bone disease or Osteosclerosis Fragilis Generalisata is a rare autosomal hereditary bone disorder of unknown aetiology. It is characterized by marked increase in bone density due to defect in remodelling caused by impaired osteoclastic function. The estimated prevalence is 1:1,00,000–500,000.²

It presents as two major forms; a clinically benign dominantly inherited (adult) form^{1–4} and a clinically malignant recessively inherited form (infantile).^{1–3} “Infantile form” is usually a severe form of disease with a diffusely sclerotic skeleton. This is characterized by normocytic anaemia with hepatosplenomegaly, optic atrophy, poor growth, frontal

bossing, pathological fractures, loss of hearing, facial palsy and increased susceptibility to infections due to granulocytopenia. Patients may present with hydrocephalous or sleep apnoea.^{1,3} Roots of teeth are often difficult to demarcate in the radiograph due to increased density of the surrounding bone. Osteomyelitis can develop secondary to tooth extraction and if untreated, patient may die within 1st decade of life due to haemorrhage, pneumonia, severe anaemia or sepsis.⁵

“Adult form” is usually diagnosed later in life and has less severe manifestations. The axial skeleton usually shows significant sclerosis, but long bones have minimal or no defects.² Bone pain is frequent in symptomatic patients. Occasionally, diagnosis may be based on the findings of the dental radiographs that show a diffuse increase

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in radiopacity of medullary bone.⁵ Patients may present with congenitally missing, delayed or unerupted malformed teeth. Erupted teeth are susceptible to caries due to reduced formation of hydroxyapatite crystals, both in enamel and dentin. Most serious complication is the increased susceptibility to osteomyelitis after tooth extraction. As the vascular supply to the jaws is compromised, avascular necrosis and infection after dental extraction may lead to osteomyelitis.²

A rare “*intermediate form*” presents during childhood with some signs and symptoms of malignant osteopetrosis.⁶ Affected individuals have a short stature, and are often asymptomatic at birth, but frequently exhibit fractures by the end of their 1st decade of life.¹ Marrow failure and hepatosplenomegaly are rare.⁴ Some patients present with cranial nerve deficits, macrocephaly, mild or moderately severe anaemia and ankylosed teeth that may predispose them to osteomyelitis.¹

This paper aims to describe clinical features, and prosthodontic management of two patients who presented with clinical features of intermediate form of osteopetrosis and missing teeth. Although conventional treatment modalities were used for the rehabilitation for the patients,

treatment planning and respective prostheses fulfilled the functional requirements of the patients.

CASE REPORTS

Case 1

A 27-year-old male patient reported to the Department of Prosthodontics, AIIMS with the chief complaints of unclear speech and inability to take liquid or solid food due to large defects in the palate. A detailed medical history revealed that patient was a diagnosed case of rare intermediate form of Osteopetrosis, associated with diminished vision, impaired hearing, and bone pain. He had a history of fracture of long bone due to which he could walk only with support. On general examination, patient had short stature, exophthalmos and depressed nasal bridge (Fig. 1). Patient had completely edentulous maxillary ridge with large antral cavities on both sides of the midline, which developed secondary to tooth extraction. Mandibular ridge was also completely edentulous except one rudimentary tooth

Fig. 1 Extra-oral view of patient, pre-treatment intra-oral view of maxillary arch, X-ray skull showing dense sclerotic bone, final prosthesis.

present in the midline. Radiographs showed diffuse sclerosis of mandibular as well as skeletal bones.

In order to improve appearance, mastication, and speech, removable definitive maxillary obturator and mandibular complete denture with relief for rudimentary teeth planned. Bilateral balanced occlusion scheme was followed for the placement of artificial teeth. Severe undercuts were blocked before making maxillary impression. Maxillary and mandibular primary impressions were made in irreversible hydrocolloid impression material using minimal pressure (Zelgan, Dentsply DeTrey GmbH, Konstanz). Mandibular secondary impression was made with zinc oxide eugenol impression paste after border moulding. Conventional procedure for fabrication of prostheses was followed. Maxillary hollow bulb obturator (with decreased bulb height) and mandibular complete denture were cured in heat cure acrylic resin (Travelon, Dentsply India Private Limited), finished, polished and delivered to the patient. Patient could not tolerate bulb with full height. So, bulb height was kept to minimum. After the final insertion, routine hygiene instructions for the dentures were given. Patient was recalled after 24 h for post-insertion adjustments and advised to remove the dentures at night to provide rest to the oral tissues. Patient was instructed to take soft or semisolid diet only, to decrease the masticatory load on the basal bone.

Case 2

A 25-year-old female patient was referred to the Department of Prosthodontics, AIIMS, with chief complaints of difficulty in chewing food due to multiple missing teeth and inadequately meeting remaining upper and lower teeth. Patient had a history of pathological fracture of long bone (femur) in 1998 for which she had to wear cast for 7 months. Patient had a history of extraction of mandibular left first molar in 2002, without any complication. Patient developed swelling over face in 2005 and underwent sequestrectomy. Teeth numbers 31, 32 and 33 were extracted during sequestrectomy. Later in 2011, resection of left mandible was done due to development of chronic suppurative osteomyelitis.

Extra-oral examination showed asymmetrical face with swelling over left side. Incomplete fusion of sagittal suture was palpable. Her maximal mouth opening was less than 1 cm. Orthopantomograph showed resected left side of mandible (Class II of Canon and Curtis classification) with missing teeth numbers 27, 31, 32, 33, 34, 35, 36, 37, 46 and 47 (Fig. 2). Maxillary and mandibular impressions were taken with irreversible hydrocolloid (Zelgan, Dentsply DeTrey GmbH, Konstanz) and casts were poured in type III gypsum product (Kalstone; Kalabhai Karson,

Mumbai, India). Bite was registered by manually guiding the mandible using minimal pressure, with silicone bite registration material (Jet bite, Coltène Whaledent, Ohio, USA) and record was transferred on the mean value articulator. Mandibular guidance appliance was fabricated in heat cure acrylic resin (Travelon, Dentsply India Ltd.) with extra row of teeth attached to maxillary removable denture. At the time of insertion, patient was advised to do lateral excursive movements for making palatal ramp with autopolymerising resin (DPI cold cure, Mumbai, India). Palatal ramp helped the patient in mastication and guide the mandible during closure (Fig. 2).

DISCUSSION

Dental abnormalities in intermediate and adult form may be attributed to the pathological changes in Osteopetrosis. Patients are susceptible to caries as not only the bone, but teeth are also hypocalcified due to imbalance in calcium and phosphorus ratio.² Constriction of canals housing neurovascular bundles that supply the teeth and the jaws, along with obliteration of marrow cavities and the dental pulp chambers, is the most likely contributing factor to bone necrosis and dental caries.⁷ Other common dental findings are congenitally missing or malformed teeth, delayed eruption, increased density of bone, thickening of lamina dura and obscured roots.² Osteomyelitis is a common complication secondary to dental caries or tooth extraction. It is a potentially severe infection that runs a protracted course, due to the accompanying severe anaemia and neutropenia.⁸

Management of patients with Osteopetrosis requires comprehensive approach as these patients have multiple problems as bone pain, fractures, impaired vision, increase chances of osteomyelitis secondary to dental treatment and neurological sequelae. Hyperbaric oxygen therapy is beneficial in the treatment of osteomyelitis as it has a bactericidal and bacteriostatic effect. It also results in improved blood supply and increased oxygen perfusion to the ischaemic area of infection.⁹

In case 1, patient presented with one rudimentary tooth in mandible and large oro-antral fistulas in maxilla, which developed secondary to tooth extraction. The rudimentary tooth was not extracted as chances of infection after extraction and fracture of jaw bone during extraction are common.⁶ Removable prosthesis without any soft liner was given to the patient as soft liners need replacement at required time intervals to maintain their effectiveness and infection free state. But it is difficult for such patients to frequently visit hospital because of bone pains and increased tendency of fracture of long bones. Removable prosthesis led to significant improvement in speech, swallowing and esthetics.

Fig. 2 OPG showing resected mandible, maxillary removable prosthesis with palatal ramp, mandibular removable prosthesis.

In case 2, patient had undergone surgery multiple times. Her mandible was deviated towards the resected side which led to asymmetric face. There are numerous methods reported in literature for reducing and minimizing mandibular deviation resulting from discontinuity defects, including mandibular guidance therapy, resection guidance restorations, or implant supported fixed prosthesis.¹⁰ But the patient just wanted a conservative treatment which could help her in chewing food. So, a maxillary removable prosthesis with palatal ramp was given. Mandibular teeth were replaced on the normal side to provide occlusion and help the patient in chewing food. Cast metal or resin based guide flange prostheses which are usually given to hemimandibulectomy patients were avoided in this patient to prevent any undue stress on the remaining bone or teeth.

CONCLUSION

Dental treatment in these patients primarily includes prevention of dental caries. Fluoride application and diet counselling is required for long term maintenance of oral

health. Special attention is to be paid if patient requires dental extraction as chances of bone infection and fracture are more. However, every effort should be made to avoid dental extraction in these cases. If patient is partially or completely edentulous only removable type of prosthesis is advisable as bones are hypocalcified and maintenance of oral hygiene is easy with removable prosthesis.

CONFLICTS OF INTEREST

All authors have none to declare.

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