Case Report

Management of a Mandibular Fracture in a Patient with Osteopetrosis: A Case Report

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Abstract

Osteopetrosis is an uncommon autosomal hereditary disorder characterized by marked increased in bone density due to defect in remodelling caused by impaired osteoclastic functioning. Surgical treatment for fractures in osteopetrotic bones is difficult due to their hardness. We report a successful surgical treatment of mandibular fracture in a 30-year-old osteopetrotic patient. Postoperatively, patient had full range of movement of mandible with good bony union and alignment.

Key Words: Osteopetrosis, mandible, fracture.

Introduction

Osteopetrosis is a rare disease characterized by systemic osteosclerosis.[1] It is a heterogenous group of inherited skeletal dysplasia's characterized by osteoclast dysfunction, impaired bone resorption and poor bone remodelling.[2] The actual incidence of osteopetrosis is unknown. However, it is thought to be 1 in 100,000 to 1 in 500,000.[3]

Two major types of osteopetrosis are recognized. These are: severe infantile autosomal recessive malignant (type I) and benign adult autosomal dominant (type II). Patients with the infantile malignant form are usually diagnosed in their first year of life and only 30% survive to the age of 6.[4] Adult form is usually diagnosed later in life and has less severe manifestations. Around 40% of patients with this type remain asymptomatic.[5] Another rare type of osteopetrosis, which is autosomal recessive is characterized by carbonic anhydrase II deficiency and is associated with renal tubular acidosis and cerebral calcification.[6]

Children who are severely affected can have cranial nerve dysfunction, and visual deficits are often evident at birth or within the first few months of life. Thrombocytopenia, anemia and infectious complications commonly cause death within the first decade. In less severe cases patients have a normal life expectancy, but the brittle bone frequently fractures, particularly in autosomal dominant osteopetrosis.[7]

Fractures in osteopetrosis can occur at any age. Because the healing response is variable, management must be adapted and individualized to address the technical challenges unique to this patient population. Operative intervention has a high rate of intraoperative and postoperative complications. [8] Surgical treatment for fractures in osteopetrotic bones is difficult as they are too hard to drill or ream. [9] A case is presented in which surgical management was carried out to fix a mandibular fracture in a patient with osteopetrosis.

Report of Case

A 30-year-old male patient who was a known case of osteoporosis, reported with a complaint of pain

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on the left side of lower jaw since 5 days. Patient gave a history of fall in the bathroom without being unconscious, vomiting or bleeding from elsewhere except oral cavity. Patient was otherwise fit and with features of osteopetrosis (Figure 1). Personal and family history was unremarkable. Extra oral examination revealed a mild swelling on the left side of mandibular body region with acute tenderness and a palpable step. There was no associated paresthesia of lip and chin region. Mouth opening was slightly restricted and intraoral examination revealed deranged occlusion on the left side. Mandibular 2nd molar was tender and infected.

Figure 1: Patient with features of osteopetrosis.

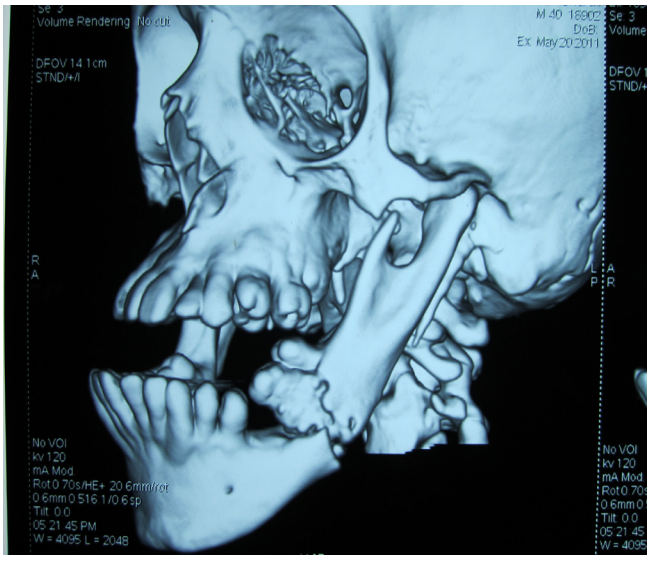

Figure 2: CT scan showing displaced fracture mandible left body region.

Patient's routine blood and urine investigations were within the normal limits. He was also investigated for serum calcium, phosphate, alkaline phosphatase levels and renal function tests, which were also normal. Radiographic examination included an OPG and CT scan (Figure 2) which showed displaced fracture mandible left body region.

Surgical Procedure

A submandibular approach was used to expose fracture fragments. Extraction of infected mandibular 2nd molar was carried out. Granulation tissue was found at the fracture site which was debrided. Fixation was done using 2.5 mm reconstruction plate with 10 mm screws (Figure 3). Closure was done using 3-0 vicryl and 5-0 prolene.

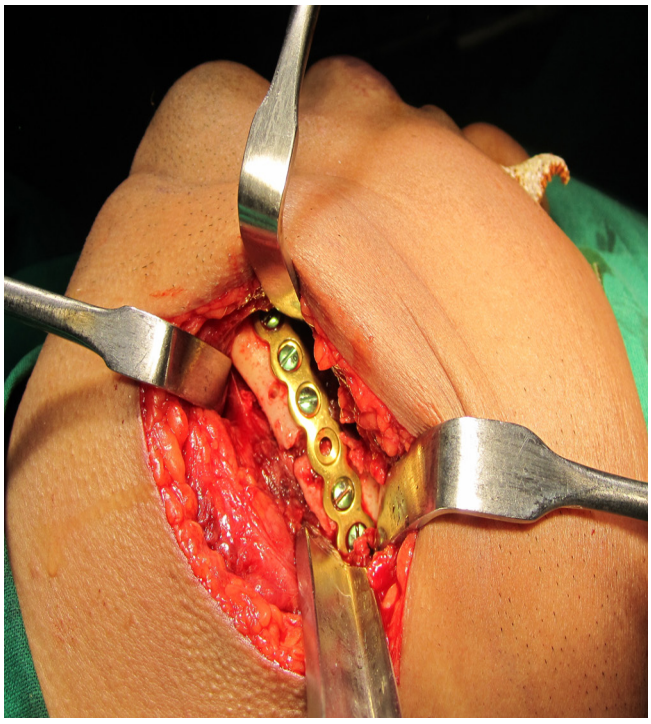

Figure 3: Mandibular fixation done using 2.5 mm reconstruction plate with 10 mm screws.

Discussion

Osteopetrosis, also known as Albers-Schonberg disease, osteopetrosis generalisata, and marble bone disease was first described in 1904 by the German radiologist Albers-chonberg.[10]

The disease represents a spectrum of clinical variants because of the heterogeneity of genetic defects resulting in osteoclast dysfunction. Failure of osteoclasts to resorb bone is thought to be the pathological defect in osteopetrosis. Research is being directed at understanding osteoclast function and developing methods that stimulate increased bone resorption or form additional sources of effective osteoclast function.[9]

Some studies have suggested gene mutation[11], carbonic anhydrase II dysfunction[12], chloride channel abnormality[13] as causative factors for various types of osteopetrosis. Classifications based on molecular events and associated physiology will undoubtedly be more precise. It is logical to expect that there is a role for hematopoietic stem-cell transplantation in patients with intrinsic osteoclast defects and severe osteopetrosis. Further investigations will facilitate correlation of the various genotypes with the clinical presentation, anticipated complications, prognosis, and expected response to treatment.[14]

Clinical manifestations in adult patients include periodic back pain, bone pain, recurrent fractures, anemia, developmental anomalies of teeth, degerative arthritis and infections.[5] Other common problems are due to compression of cranial nerves causing blindness, hearing and vision loss and facial nerve palsy.[15]

Radiographic features include increased density of the entire skeleton due to a lack of cortical endosteal margins and failure of bone remodelling.[16] It causes hard and brittle marble bone fracturing easily. Most of these fractures can be treated conservatively.

Operative intervention when needed presents with unique technical challenges. In this case an extra oral submandibular approach was used to provide better soft tissue coverage after fixation particularly in an osteopetrotic patient.

While osteopetrotic hard bone may be penetrated with a drill bit; high friction and prolonged drilling can make the drill bit blunt. The heat generated can cause bone necrosis and break the drill bit. Besides this, brittleness of bones can cause intraoperative fractures. Due to the difficulties during the operation, the operative time may be prolonged thereby increasing the risk of postoperative infection. There is also a risk of delay in consolidation and nonunion owning to impaired bone remodelling. In order to overcome the technical difficulties regarding drilling and reaming of hard sclerotic bones, recommendations have been made to use high speed electric drill bits, frequently cooling them and using the graduated drill bit system to overcome drill breakage and overheating.[17]

While drilling, the flutes of the drill bit are frequently filled with dense accumulated bone swarf, which reduces its effectiveness. Frequent exchange of drill bits, cleaning of their flutes from the filling hard bone, constant saline cooling during drilling is obligatory.[18]

In this case we encountered difficulty while drilling the bone. High speed stainless steel drill bit of 2.0 mm was used with copious irrigation. To overcome the resistance in penetrating the bone and the drill bit was changed twice. The swarf was removed from the flutes of drill bit.

Titanium reconstruction plate (2.5 mm) and screws were used to fix the fracture fragments as this provided rigid internal fixation (Figure 4). Postoperatively the patient had uneventful recovery and has been followed up for one and a half year There has been no episode of any further swelling, pain or infection. Patient had mild paresthesia in the immediate post-operative period but this gradually recovered. Mandibular movements were in normal in the follow-up period.

Figure 4: Postoperative OPG.

Key Points

- There is not much literature available regarding treatment of mandibular fracture by open reduction and internal fixation in osteopetrotic mandible but there are reported cases of treatment of long bone fractures with plates and screws with uneventful results.
- Thus, surgery can be difficult in terms of drilling. Adequate cooling during drilling is necessary to avoid postoperative necrosis of the bone.
- Open reduction and internal fixation in osteopetrotic mandible is not contraindicated provided good preoperative planning is done anticipating the possible technical difficulties. The prolonged intraoperative and postoperative course and risk of infection should be included in the informed consent.

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