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CASE REPORT
Craniofacial/Pediatric

Surgical Treatment of a Bilateral Mandibular Fracture in a Patient with Osteogenesis Imperfecta Type III

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Summary: Osteogenesis imperfecta is a group of genetic disorders affecting skeletal and connective tissue that commonly presents with a history of multiple fractures. In particular, Osteogenesis imperfecta type III is associated with severe bone deformities, specifically in the mandible with deformities such as mandibular overjet and cross-bite. We report a case of a bilateral mandibular fracture in a young adult diagnosed with Osteogenesis imperfecta type III, managed surgically with open reduction internal fixation. *(Plast Reconstr Surg Glob Open 2021;9:e3702; doi: 10.1097/GOX.0000000000003702; Published online 19 July 2021.)*

Osteogenesis imperfecta (OI) is a genetic disorder of skeletal and connective tissue that is characterized by bone fragility and low bone mass, often referred to as "brittle bone disease." It is estimated that the incidence of OI is approximately one per 20,000 births, and an estimated 20,000 to 50,000 people in the United States have the condition.[1] Patients with OI may present with extra-skeletal clinical features such as blue sclera, dentinogenesis imperfecta, hearing loss, ligament laxity, short stature, easy bruising, excessive fractures of the extremities, and uncommonly, fractures of the facial bones.[2,3]

The broad spectrum of OI has resulted in comprehensive classification systems that were first proposed by Sillence et al. OI type I is associated with mild to no bone deformities. OI type II is associated with severe bone deformities and perinatal death. OI type III is associated with growth retardation and severely progressing bone deformities. OI type IV is associated with mild growth retardation and moderate bone deformities.[4] Bisphosphonates are the main pharmacologic treatment modality as they inhibit bone resorption and turnover to prevent fracture risk, although not specifically approved for treatment of OI.[5] In addition, studies have shown that patients with OI commonly have several deviations in the orofacial area. Specifically, one recent study found that moderate–severe cases (types III and IV) have mandibular overjet and posterior cross-bite.[6] Other studies indicate that class III malocclusion with mandibular overjet is prevalent in OI type III specifically.[7,8]

To the best of our knowledge, mandibular fractures are uncommon in OI patients, and the temporomandibular disorders have been attributed to OI type III, adding to the complexity of surgical management. We report a bilateral mandibular angle fracture after a fall in a young adult patient with OI type III.

CASE REPORT

A 19-year-old man reported to the emergency department for evaluation of a bilateral mandibular fracture. Medical history revealed that the patient was diagnosed with type III OI and had an extensive fracture history. Of note, the patient has no history of facial fractures and was not on any medications, including bisphosphonates.

The patient reported that he fell down, striking his jaw on the ground with immediate pain and malocclusion. Clinical examination showed oral bleeding from tooth #32, as well as facial swelling and trismus. A panoramic radiograph revealed bilateral mandibular fractures (Fig. 1).

Open reduction internal fixation (ORIF) was recommended, and all benefits, risks, and alternatives were discussed with the patient. The procedure began first by placing the patient into intermaxillary fixation to obtain the best occlusion based on the wear facets. An external approach was chosen to allow for exposure and placement of the reconstruction bars. The fractures were isolated appropriately, and reduction forceps were then placed along the inferior margin of the mandible and reduction was attempted. However, the fracture was unable to be reduced due to the

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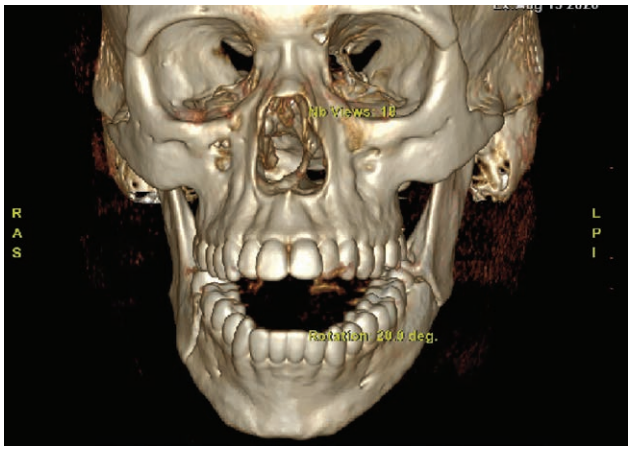

Fig. 1. 3D CT reconstruction (anterior view), bilateral mandibular angle fractures. Left mandibular angle fracture, closed, unfavorable, noncomminuted, minimally displaced. Right mandibular body fracture, closed, unfavorable, comminuted, nondisplaced.

intermaxillary fixation that was placed before the external approaches. The intermaxillary fixation was then loosened and the fracture was able to be mobilized. Once both retraction forceps were situated, both fractures were plated with a 2.0-mm, four-hole reconstruction bar and 10.0-mm bone screws. The mandible was stable and manual palpation showed minimal step-off on the inferior of both cortices as well as the lingual surface of both fractures. After ORIF, the wear facets did not align, and there was an open bite deformity. Another facial trauma surgeon was consulted to evaluate the reduction. It was decided to discontinue further surgical treatment and get a repeated CT with 3D reconstruction to evaluate the reduction. Based on the patient's history of type III OI, it was felt this was the best option to limit the risk of further fractures on an already stable and reduced mandible. The patient's mother also stated that he had a baseline open bite deformity. (Figs. 2–4)

The patient had an uncomplicated postoperative period and was placed on a soft diet for 4–6 weeks. At 6 weeks follow-up, he stated his occlusion was normal and he was satisfied with his result.

Fig. 2. Postoperative reconstruction (anterior view) showing CT 3D reconstruction plate fixation.

Fig. 3. Postoperative reconstruction (right lateral view) showing CT 3D reconstruction plate fixation.

DISCUSSION

Minimal studies have investigated the temporomandibular disorders of OI patients. As described by Sillence et al, there are four major types of OI based on the severity and progression. Specifically, studies have shown that patients with OI type III have a higher likelihood of mandibular overjet and class III malocclusion. This is consistent with our patient before his mandibular trauma and raised concern following successful reduction.[7,8]

During mandibular trauma and the decision to undergo ORIF, it is of utmost importance to ensure the fracture is successfully reduced and to re-establish the patient's pre-injury dental occlusion.[9] When reducing fractures in patients with OI type III, it is recommended to use large reconstruction bars. However, it was noted that the patient's occlusion was not in-line and there was an open bite deformity. This raised concern by the surgical team, as malocclusion can be a significant postoperative complication. It is most often due to technical error and if not recognized intraoperatively, the patient will need to return to the operating room for corrective surgery.[9] Because this was the first encounter with the surgical team

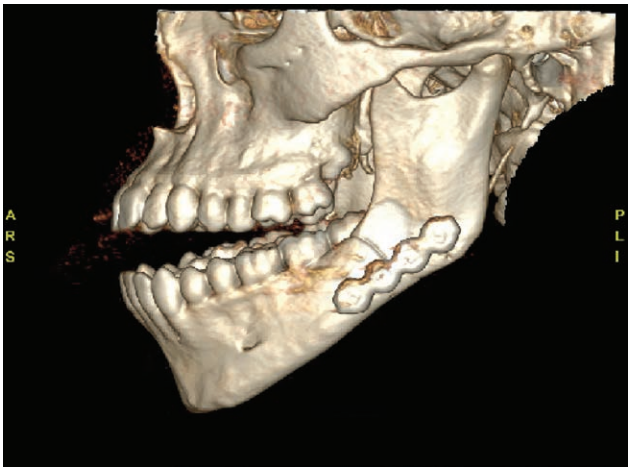

Fig. 4. Postoperative reconstruction (Left lateral view) showing CT 3D reconstruction plate fixation.

and previous medical records made no mention of his normal mandibular alignment, the patient's family was consulted. The patient's mother reassured the team that his malocclusion and open bite were both normal before his trauma. After undergoing ORIF and an uncomplicated postoperative course, the patient did in fact confirm that this was his normal alignment.

CONCLUSIONS

Meticulous care is necessary for mandibular fractures in patients diagnosed with OI, even when the nature of the fracture and surgical intervention can be successfully achieved with adequate fixation and reduction. It is important to remember when evaluating OI patients with mandible fractures, that they commonly have temporomandibular disorders and malocclusion deformities. This may provide a challenge for the surgeon to adequately perform an ORIF operation; however, this may be a normal dysfunction and should be considered.

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