Surgery for clavicular and humeral fractures in an osteopetrotic patient: a case report

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ABSTRACT

Osteopetrosis is a rare disease characterised by generalised sclerosis of the bone. Surgical treatment for fractures in osteopetrotic bones is difficult due to their hardness. We report successful surgical treatment of humeral and clavicular fractures in a 30-year-old osteopetrotic patient with severe multiple trauma. Two years after surgery, the patient had a full range of movement at the shoulder and elbow, with good bone union and alignment.

Key words: fracture fixation, internal; fracture fixation, intramedullary; osteopetrosis

INTRODUCTION

Osteopetrosis is a rare disease characterised by generalised sclerosis of the bone. Surgical treatment for fractures in osteopetrotic bones is difficult as they are too hard to drill or ream. Surgical treatment for fractures in osteopetrotic bones is difficult due to their hardness. We report successful surgical treatment of humeral and clavicular fractures in a 30-year-old osteopetrotic patient with severe multiple trauma. Two years after surgery, the patient had a full range of movement at the shoulder and elbow, with good bone union and alignment.

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CASE REPORT

In June 2003, a 30-year-old man was injured in a motorcycle accident and presented to our hospital. He was conscious but vomiting and dyspnoeic. A computed tomographic scan of the brain revealed an acute intracranial epidural haematoma that was evacuated immediately. A haemothorax and multiple right rib fractures were treated conservatively. Sclerotic changes in the lumbar spine and pelvis were detected during examination of the abdominal and thoracic radiographs (Fig. 1). The lumbar vertebrae showed thick dense endplates on the upper and lower borders of the vertebral bodies, with normal density in the middle, known as a 'Rugger Jersey spine'. Sclerosis at the sacroiliac joints and iliac acetabula
strongly suggested osteopetrosis.

The patient’s condition was stabilised within one week and he was transferred to the orthopaedic department. He complained of severe pain and swelling in the right shoulder and brachial area. There were no neurovascular deficits in his right arm. Radiographic examinations revealed fractures of the clavicle and humerus (Fig. 2). The clavicle was fractured at the distal third and the medial segment was displaced due to rupture of the coracoclavicular ligaments—a Neer type-II distal clavicle fracture. The humerus was fractured transversely at the middle of the shaft. Both fractures required surgery.

The distal clavicular fracture was treated first with the patient in a supine position under general anaesthesia. A skin incision was made to expose the fracture site. The trapezoid ligament remained attached to the distal clavicle, but the conoid ligament was torn together with the comminuted bone fragmentation. The clavicle was reduced and fixed with an acromioclavicular plate (BEST Medical Co., Japan) and reinforced with cerclage wiring (Fig. 3).

The humeral shaft fracture was then treated. A skin incision was made on the lateral shoulder and the deltoid and rotator cuff were split. An additional approach was made to the posterior humerus to protect the radial nerve. Open reduction and internal fixation using an interlocking intramedullary nail (Humeral Nail System, ACE Co., US) were performed.
The entire procedure—inserting the guide wire, reaming the bone cavity, and drilling and screwing the bone cortex—was difficult because of the very hard and thick bone cortex. Nonetheless, sufficient fixation and anatomical alignment of both the clavicle and humerus were achieved.

Immediately after surgery, shoulder sling exercises were started, initially limited to a maximum elevation of 90°. Three months postoperatively, distal clavicle bone union and abundant callus formation was confirmed radiologically so the acromioclavicular plate was removed. Unrestricted range-of-movement shoulder exercises were allowed. Two years after surgery, the humerus and clavicle showed good union and alignment (Fig. 4). The patient had returned to work and had a full range of movement at the shoulder and elbow.

DISCUSSION

Osteopetrosis, also called marble bone disease, was first described by Heinrich Albers-Schonberg. It is a rare developmental bone disorder characterised by generalised sclerosis of bone. Radiographic features include increased density of the entire skeleton due to a lack of cortical endosteal margins and failure of bone remodelling. Osteopetrosis varies in severity depending on the time of recognition and is classified into either an infantile malignant type or an adult benign type. The infantile malignant type is autosomally recessive, has an onset at, or shortly after, birth and is lethal. By contrast, the adult benign type, which is autosomally dominant, has its onset in later childhood and is usually discovered accidentally during a radiographic examination for another condition. Based on radiographs, autosomal dominant osteopetrosis is classified into 2 subtypes with different clinical, biochemical, and histological manifestations. Type I is characterised by massive sclerosis of the skull and thickening of the cranial vault, while in type II the sclerosis of the skull is most striking at the base. In type II, particularly, the spine shows thick dense endplates at the upper and lower borders of the vertebral bodies, with a normal appearance in the middle third known as a ‘Rugger Jersey spine’. Sclerosis is also seen in the pelvis and fingers. Our patient was diagnosed as having sporadic autosomal dominant type-II osteopetrosis.

The Neer type-II distal clavicle fracture is characterised by disruption of the ligamentous attachment from the proximal fragment. It has a high rate of non-union and prolonged pain and should be treated case-by-case. Some authors recommend internal fixation with Kirschner wires, Kirschner wires with tension bands, cerclage wires, and screws for severe displacement or no bone contact. A clavicular hook plate fixation system has advantages over Kirschner wires for treatment of the type-II distal clavicle fracture as Kirschner wires may result in migration and loss of reduction, even though shoulder movement may be limited prior to clavicular hook plate removal.

For humeral shaft fractures, non-surgical treatments (such as hanging cast, U-shaped coaption splint, and functional orthosis) have yielded satisfactory outcomes. Nonetheless, cases in which an acceptable reduction cannot be maintained should be treated surgically. Our patient had multiple trauma and it was not possible to maintain a closed reduction. Surgical treatment of our adult patient with benign osteopetrosis achieved good results despite the difficulties of drilling and reaming the bones.
REFERENCES