Case report

Accordion Maneuver with Modified Ilizarov Frame for treatment of recalcitrant nonunion of shaft of femur in a patient with Pycnodysostosis

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ABSTRACT

Accordion Maneuver is a strong “Bloodless Tool” in the Ilizarov technology, that helps to bring about healing of nonunion of fractures of long bones. A 50 years old male, a rare case of Pycnodysostosis, presented to us with recalcitrant nonunion of the fracture shaft of the femur after two failed surgical attempts, with a broken plate in the last attempt. We treated this very difficult nonunion by Ilizarov monofocal osteosynthesis with Accordion Maneuver that finally yielded union. This appears to be the only case report of treating such a case with Accordion Maneuver.

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1. Introduction

Accordion Maneuver (AM) is a “Bloodless Tool” to stimulate bone healing in nonunion of fractures of the long bones as was described by Professor G.A. Ilizarov. It comprises of alternate compression and distraction at the fracture site that converts the scar tissue at the fracture site into tissues capable of neo-osteogenesis.¹ We are discussing the role of this tool while treating a recalcitrant nonunion of fracture shaft of the femur by modified Ilizarov system in a rare case of Pycnodysostosis who underwent multiple operations earlier for the same fracture. To our knowledge, this is the only case report of such a procedure in this rare condition.

2. Case report

A 50 years old male, reported after two failed attempts to treat a fracture shaft of the right femur in its middle third. He was earlier operated successively with open reduction and internal fixation by K-nail and then with plate and screw. Clinical assessment revealed that he was four feet three inches tall with receding jaws,

micrognathia and prominent nose. His hands were short with stubby fingers (Fig. 1). Similar clinical findings were found in his younger sibling. On local examination, a diagnosis of nonunion was made as there was frank abnormal mobility at the fracture site without pain. X-ray revealed dense bone with no attempt of callus formation with a broken plate (Fig. 2). It was presumed that, multiple operations led to poor vascularity at the site and the stability provided by the earlier implants was not adequate to maintain the alignment till the revascularization process was complete. Since both intramedullary and extramedullary devices failed in this case, we looked for the way by which we could give the best chance of union. Though Interlocking Nail with bone grafting could have been used in this situation, we wanted to obviate the need for bone grafting, which would prevent additional donor site morbidities specially in diseased bone. Therefore we decided to treat the patient with a Modified Ilizarov Frame and an Accordion Maneuver was also planned to be a part of the treatment protocol. Patient was operated and the fracture site was approached through the old scar. The broken implants were removed, the margins were freshened and a modified Ilizarov Ring Fixator was mounted. The system consisted of an Italian arch and three rings; the arch and a ring were anchored to the proximal fragment through 4.5 mm Schanz screws while the distal fragment was anchored with Kirshner wires through another two rings (Fig. 3). Accordion Maneuver was started after three weeks of operation and was continued for three cycles. Compression was started first. Each cycle consisted of compression for seven days, at

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Fig. 1. Abnormal clinical features of Pycnodysostosis.

the rate of 0.25 mm in 12 h, followed by distraction for seven days and rest for 3–5 days. After three such cycles, compression was started at the rate of 0.25 mm on every 3rd day till trans-cortical bone growth could be seen across the fracture site in X-ray. Dynamisation was done to confirm the status of healing. It involved loosening of one nut of a threaded rod and continuing weight bearing to see if there was any pain at the fracture site. Nuts in the opposite rods of same end were loosened sequentially every seven days so that at end of four weeks all the nuts in four rods were loose. Ultimately fracture united at about six months (Fig. 4).

There was no re-fracture when the patient visited us at the third year after implant removal.

3. Discussion

Pycnodysostosis is a rare autosomal recessive skeletal dysplasia. It is caused by mutation in the gene that codes the lysosomal enzyme cathepsin K on chromosome 1q21, responsible for degradation of collagen type 1.² This enzyme is important for activity of osteoclasts. Thus, in individuals with Pycnodysostosis remodeling of bone is affected and bones become abnormally sclerotic, dense and brittle.² It is often confused with osteopetrosis because of increased density of bone and also with cleidocranial dysostosis because of skull and clavicle abnormalities. Dwarfism is constant with height not more than five feet. The skull appears wide, the jaws are receding, the hands are short and with stubby fingers. Radiologically, there is failure of closure of cranial sutures and with wormian bones, hypoplastic mandible ramus, acro-osteolysis of terminal phalanges, clavicles poorly formed with absence of acromial ends. The entire skeleton appears denser than

Fig. 2. Failed plate osteosynthesis.

Fig. 3. Mounting of Ilizarov Frame.

Fig. 4. Union of the fracture.

normal.³ There is osteoclastic dysfunction and few workers have studied the condition extensively in recent times.^{4–6}

We diagnosed the patient to suffer from Pycnodysostosis based on the clinical and radiological findings. The probable cause of failures of the previous operations could be inadequate stability provided by the implants and may have been contributed by the disease (though we cannot be certain as previous X-rays were not available).

Ilizarov proved that controlled process of distraction helped in creation of mechano-biological conditions (tissue strain) that stimulate new bone formation mostly by intra-membranous ossification.⁷ Claes concluded that temporary cyclic distraction and compression could enhance better bone healing by producing early increased bone volume that had higher bending rigidity.⁸

Transformational Osteogenesis is a technique in Ilizarov system used in nonunion to stimulate healing. The tools used in this method are compression and distraction depending on the type of tissues between the fragments. There is transformation of collagen tissues between the fracture surfaces at nonunion site into tissues capable of transforming into bone. This is accomplished by what is called Accordion Maneuver as it comprises of alternate compression and distraction like the musical instrument so named. The selection of whether to start with compression first or distraction first depends on the quality of tissue anticipated between the fragments at the nonunion site. In stiff (hypertrophic) nonunion, fibrocartilage and or synovial tissue is anticipated and therefore

distraction is started first to initiate neo-osteogenesis followed by compression to consolidate the new bone so formed. On the other hand, in mobile (atrophic) nonunion, fibrous tissue is anticipated at the nonunion site and therefore compression is done first to crush the tissue to produce an environment of inflammation that is conducive for neogenesis followed by distraction. Distraction creates columnar fibro-vascular tissues that arise from the crushed tissue on fracture surface. Repeated distraction stimulates production of osteoblast and helps collagen bundles to consolidate within a bony matrix.⁹

Generally, the suggested protocol for this maneuver in nonunion of fractures is compression for 10 days, followed by distraction up to 10–20 mm, at the rate of 0.25 mm twice a day, which is followed by rest for 7–10 days; and then slow compression again up to 7–10 mm. After a rest period for 5–7 days, distraction is performed for second time.¹ We modified the protocol of AM according to our experience.

Although a bifocal osteosynthesis with compression at the fracture and distraction through a corticotomy would have been ideal for our case as suggested by ASAMI group,¹⁰ a monofocal osteosynthesis was planned and executed as there was fear that a corticotomy might lead to another nonunion at the corticotomy site in addition to existing nonunion. We therefore relied solely on the capacity of AM to stimulate heal process in this case of recalcitrant nonunion.

To conclude, Accordion Maneuver is an excellent tool which can stimulate healing process in recalcitrant nonunion even without the need for corticotomy. A modified protocol, by reducing some days in between, can yield the same result as with original protocol suggested by the masters.

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Conflicts of interest

The authors have none to declare.

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