

A challenging case of Pelvi-Acetabular fracture in a patient with Achondroplasia .

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ABSTRACT

Achondroplasia is characterized by dwarfism and typical changes in the size and shape of bones particularly of the pelvis. Additional anatomical features and presence of co-existing medical problems, poses significant anaesthetic problems. Pelvic and acetabular fractures in achondroplasia are rare and there is a scarcity of literature about management of these fractures. We report a case of 55 years old lady with achondroplasia who sustained pelvi-acetabular fracture that was successfully managed by two staged surgery, to highlight the orthopaedic and anaesthetic challenges.

Key words: Achondroplasia, pelvic, acetabulum, fracture

INTRODUCTION

Fractures in a patient with achondroplasia pose orthopaedic and anaesthetic challenges for management. Various bony and soft tissue abnormalities are well documented, which results in difficulty of access to the operative field and need for special instrumentations and implants to fix the fractures.^{1,2} Anaesthetic challenges include long hours of operation, difficulty in positioning and intubation, increased blood loss, and restrictive lung disease.³ In this case report we highlight the challenges of surgical management of pelvi-acetabular fracture in a 55 years old lady with achondroplasia.

CASE REPORT

A 55-year-old lady with achondroplasia sustained a pelvi-acetabular fracture following a road traffic accident. She was about 30 kg in

weight and 120 cm in height. She was an asthmatic on bronchodilator inhalers. She was haemodynamically stable. Radiographs of the pelvis and acetabulum and CT scan of the pelvis revealed comminuted fracture of the right ilium extending to the acetabulum, undisplaced fracture of the left ilium, bilateral superior pubic ramii and left inferior pubic ramus fracture and diastasis of the left sacroiliac joint (Figs. 1, 2).

Her blood investigations were normal and an echocardiogram showed an ejection fraction of 67%, normal valves and chamber dimensions with minimal left ventricular hypertrophy and mild diastolic dysfunction. There were no features of pulmonary hypertension or cor pulmonale.

Anaesthetic assessment showed minimal restriction of neck movements, but adequate mouth opening with oropharyngeal class III: visualization of the soft palate and

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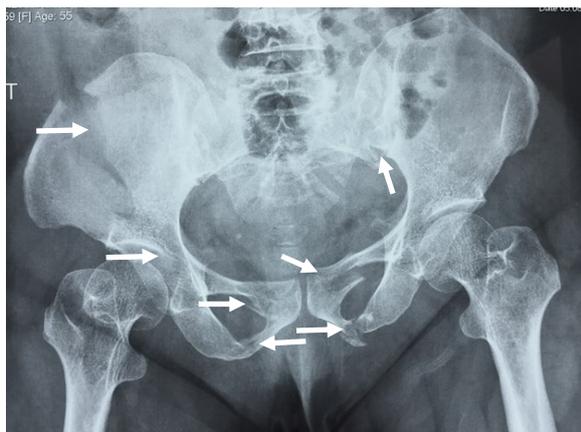


Fig. 1: Pre-operative radiograph (White solid arrows indicating points of fracture of the pelvis)

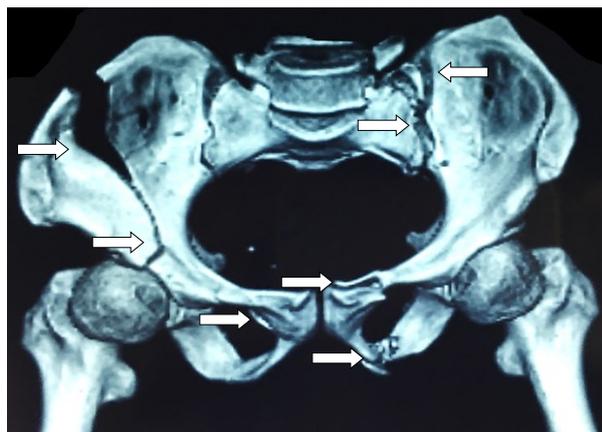


Fig. 2: Pre-operative 3-D CT reconstruction (White solid arrows indicating points of fracture of the pelvis).

the base of uvula according to the Mallampati score. There were no features of obstructive sleep apnoea. She was noted to have ASA III according to the American Society of Anesthesiologists (ASA) physical status classification evaluation suggesting severe systemic disease that limits activity but is not incapacitating. The patient was also counselled by the anaesthetists about the various procedures involved before the first stage surgery.

She was scheduled for an open reduction and internal fixation of the acetabulum and ilium as well as the contralateral sacroiliac joint, on a semi-urgent basis in stages anticipating intra-operative difficulties and challenges from prolonged anaesthesia after discussion with the anaesthetist.

In the first stage, adequate venous access and non-invasive monitoring was first established. For anaesthesia, fiberoptic intubation was performed orally using a 6.0 cuffed PVC endotracheal tube taking extreme care to minimise all neck movements and keeping difficult airway cart as standby. Surgery was commenced after adequate positioning and ensuring padding. An anterior ili-inguinal approach was used with the patient in the supine position. For fixation of the anterior column fracture, a 6-hole well contoured AO reconstruction plate was used to span over the endopelvic surface of acetabu-

lum (quadrilateral plate), and fixed to the pubic ramus. A second 6-hole AO reconstruction plate was used to span the iliac crest bridging the two halves of the anterior wing of Ilium that was separated by the injury. The duration of surgery was 3 hours 45 minutes. After routine reversal of anaesthesia patient was shifted to intensive care unit for monitoring.

After a gap of 1 week, in the second stage, the sacroiliac disruption was fixed with percutaneous single cancellous AO 7.3 mm half threaded screw with washer to effect compression of iliosacral disruption. This procedure was also performed under general anaesthesia with due care as during the first stage and lasted 1 hour and 15 minutes.

Post-operative radiographs were satisfactory (Fig. 3). She was started on chest and upper and lower limb exercises in bed. In view of her osteoporotic bones, weight bearing was delayed for 6 weeks and she was allowed ambulation in a wheel chair. At discharge, she was commenced on anti-resorptive therapy using alendronate, calcitriol and calcium lactate.

At last followup, 12 months after surgery, the patient was able to walk full weight bearing without limp. Follow up radiographs revealed good healing of fractures with maintained reduction and implant position.



Fig. 3: Post-operative radiograph showing plates and screws using for internal fixation of the fractured components of the pelvis.

Discussion

There is paucity of literature about management of fractures in patients with achondroplasia, which presents both orthopaedic and anaesthetic challenges.

Based on the study of literature and analysis of the imaging studies, the following issues related to surgery were anticipated and discussed. The deformed and small bones, restricted exposure, osteoporotic bones and need for specialized instrumentation and implants.

The pelvis and hip differs from normal patients in the horizontal acetabular roof (decreased acetabular angle), small squared iliac wings, small trident pelvis, champagne glass type pelvic inlet, short sacroiliac notches and hip flexion contractures.^{1,2} This may result in the requirement for modifications and possible compromises in the fixation.⁴ Anatomical peculiarities and possible joint contractures may make positioning difficult, and special care must be taken to position and pad pressure points adequately. However, in our case we did not have to use any special approach or implants. Low bone density has also been documented in a recent study on patients with achondroplasia.⁵ Due to lack of AO locking reconstruction plates, which would have been ideal for this case, regular plates

were used. Owing to the smaller size and altered shape of the sacral promontory, fixation of the left sacro-iliac joint was done using a single screw which could not cross the midline as is generally practiced.

In the management of a patient with achondroplasia and distal femur fracture, Murphy *et al* have highlighted the problem of abnormal anatomy and size of bones which makes use of routine implants difficult.⁴ They have also referred to poor bone quality in such patients which can compromise the stability of the fixation.

From anaesthesia consideration, achondroplastics have facial features that may make airway management problematic. In addition, there is limited flexibility of the atlanto-occipital joint with risk of foramen magnum stenosis and cervical instability.³ Due to the above issues, it was decided to secure the airway by fiberoptic intubation, minimising neck movements. Selecting the size of the endotracheal tube in achondroplastics must be based on body weight rather than age. Hence a size 6 endotracheal tube was used. Although these patients are described to be anxious, there is a possibility of obstructive sleep apnoea in addition to airway difficulties.⁶ Hence we chose to allay the patient's anxiety by explanation and reassurance and not sedatives or anxiolytics.

In conclusion, patients with achondroplasia are affected by numerous medical comorbidities and this combined with the disproportionate bony abnormalities can raise many challenges in surgical management of fractures. Adequate pre-operative workup, careful planning with multi-disciplinary approach for perioperative care can help in avoiding complications.

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