# AN UNUSUAL CASE OF ATLANTO AXIAL INSTABLITY IN A PATIENT OF SKELETAL DYSPLASIA WITH BILATERAL TOTAL HIP REPLACEMENT, SUGGESTING NEED FOR CAREFUL SPINE EVALUATION

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#### ABSTRACT

Skeletal dysplasia's are a heterogeneous group of disorders that have in common a defect in bone formation or remodeling, thus leading to abnormal biomechanics and various spinal and joint manifestations. A 25 year old female presented to us after bilateral Total Hip Replacement along with bilateral adductor tenotomies with severe hamstring tightness. Our suspicion of atlanto-axial instability was confirmed after radiological investigation in form of Dynamic X-rays and Magnetic Resonance Imaging of cervical spine with cranio-vertebral junction. Posterior Atlanto-Axial fixation and fusion was done followed by

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physiotherapy. Lhermitte's sign disappeared immediately following surgery. Intermittent stretching of hamstring followed by skin traction with 2 kilo weight was done. Once full extension was achieved, it was maintained in extension brace. At last follow-up of 1 year patient spasticity has improved to scale 2 and is able to walk with stick. C1-2 instability as part of Cranio-vertebtral junction anomaly is most common spinal manifestation in patient with spinal dysplasia leading to spastic quadruparesis. Screening of Craniovertebral junction anomaly must be done in all patients of Skeletal Dysplasia.

KEYWORDS: Skeletal Dysplasia, Cranio-vertebral junction, Atlanto-axial instability, Screening.

### **INTRODUCTION**

Skeletal dysplasias are a heterogeneous group of disorders that have in common a defect in bone formation or remodeling, thus leading to abnormal biomechanics and various spinal and joint manifestations. Atlantoaxial instability, is radiologically identified increased mobility or laxity between the body of the first cervical vertebra (atlas) and the odontoid process of the second cervical vertebra (axis). The subluxation can be anterior, posterior, or lateral, and symptoms occur as a result of cervical cord impingement. This instability can originate from congenital conditions, but in adults, it is primarily seen in the setting of acute trauma or degenerative changes due to the inflammatory pannus of rheumatoid arthritis (RA). Infection has been found to be an additional cause of instability, with the rich arterial supply and venous plexus in this region of the body providing a route for infectious sequelae.

#### **CASE REPORT**

A 25 year old short stature female presented to us with progressive bilateral hip arthritis due to underlying

severe bilateral hip dysplasia, patient was in severe hip pain and wheelchair bound. Due to her severe pain and advanced bilateral hip arthritis (Fig 1), decision was made to perform a bilateral total hip replacement surgeries in 2 stages in a 3 week interval period. After the replacement surgeries (Fig 2), she was pain free and was mobilized with walker, though there was mild hamstring stiffness present. She presented to us again in 4 weeks with a acute onset of paraesthesiae in the fingers of both hands. This began suddenly and she subsequently developed a progressive weakness in all four limbs until she was unable to walk or stand. On careful neurological evaluation it was found that patient had spastic quadriparesis with restricted neck range of motion and positive Lherrmitte's sign (sudden sensation resembling an electric shock that passes down the back of the neck and into the whole spine and may then radiate out into arms and legs. It is usually triggered by neck flexion). Sensory examination was normal. Patient had 60 to 120 degree range of motion in both knee with spasticity of scale 4 on modified Ashworth Scale. The suspicion of Atlantoaxial instability (AAI) was confirmed after radiological

investigations in form of Dynamic X-rays and Magnetic Resonance Imaging of cervical spine with Cranio-Vertebral (CV) junction scanning (Fig 3-4). Posterior Atlanto-Axial fixation and fusion was done followed by physiotherapy (1-2). (Fig 5)



Fig 1: Preop X ray Before Bilateral Hip Replacement



Fig 2: Postop X-rays After Bilateral Total Hip Arthroplasty



Fig 3: X-rays Showing C1-C2 Instability ERA'S JOURNAL OF MEDICAL RESEARCH, VOL.6 NO.2



Fig 4a: CT Scan and MRI



Fig 4b: Axial Images



Fig 5: Postop X-rays After Pedicle Screws Fixation

Lhermitte's sign disappeared immediately following surgery. Intermittent stretching of hamstring followed by skin traction with 2 kilo weight was done. Once full extension was achieved, it was maintained in extension brace. Full extension of bilateral knee was attained after 3 weeks and patient was mobilized out of bed with walker and brace. At last follow-up of 2 years patient spasticity has improved to scale 2 and is able to walk with stick (3-4).

## DISCUSSION

Atlantoaxial instability as part of Cranio-vertebral junction anomaly is the most common spinal manifestation in patients with spinal dysplasia leading to spastic quadruparesis (5). Screening of craniovertebral junction anomaly must be done in all patients of Skeletal Dysplasia (6). Some authors also suggest routine investigation in the absence of symptoms and even prophylactic cervical fusion at an early age in high risk patients (7). Patients with short stature require specific precautions for surgery including the cervical fusion required in our patient to prevent her imminent quadraplegia. Neck flexion or extension should be avoided, which can be achieved by cervical immobilisation in a collar. The patient can also be intubated for surgery using a nasotracheal fibre endoscope (8). In the event of emergency surgery the anaesthetist must presume that the patient with short stature has an unstable cervical spine. At present the optimal surgical management of atlantoaxial instabilty is either posterior atlantoaxial fusion or fusion of the posterior axial arch to the occiput.

## CONCLUSION

Atlantoaxial instabilty must be suspected in patients with abnormalities of skeletal development presenting with limb weakness, spasticity or paraesthesiae. Care should be taken in all patients of short stature requiring intubation for general anaesthesia.

Atlantoaxial instability may result from conditions other than rheumatoid disease and the seronegative spondyloarthropathies.

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