Atlanto-Occipital Dislocation in Rickets: Letter to Editor

A three year old boy fell down while playing and could not move any of his four limbs. There was no loss of consciousness or no respiratory difficulty. He had no sphincter disturbances. The child was being treated for nutritional rickets by vitamin D analogues and calcium supplements. He was initially managed at a local hospital and referred to us after a week. Examination revealed normal vital parameters and no external injuries. Neurological examination showed normal higher mental functions, right sided Horner’s syndrome and flaccidity of all four limbs. Power in the upper limbs was 2/5 proximally and 0/5 distally. In the lower limbs, power was 3/5. Sensations were impaired below neck. Plantar reflexes were extensor bilaterally. Cremasteric and abdominal reflexes were absent. Deep tendon jerks were absent in upper limbs and exaggerated in lower limbs. Routine hematological and biochemical investigations were normal. X-ray cervical spine and a tomogram showed posteriorly subluxated atlas and odontoid process and posterior atlanto-occipital dislocation (Fig. la). X-ray of right wrist showed zone of preparatory calcification and calcification of rachitic metaphysis (Fig. lb). He was managed initially with steroids and strict immobilisation. Traction could not be applied due to soft bones in rickets. He gradually improved in motor power to 3/5 in upper limbs and 4/5 in lower limbs. He underwent C1 posterior arch excision and laminectomy of C2 vertebra. There was kink between the foramen magnum rim and posterior arch of atlas. Fusion was not done as patient was considered very young and it was planned to do it at a later date if required. Postoperative period was uneventful except for occipital pressure sore. He gradually improved and at one year he had only mild right hemiparesis.

Rickets affects all the stages of bone formation i.e. proliferation, maturation and destruction. This results in defective growth of bone due to retardation or suppression of normal growth of epiphyseal cartilage and normal calcification of the matrix. The bones are softened and rafified. The cortical bone is readily distorted by stress. Relaxation of the ligaments results in deformities such as kyphosis and scoliosis.1 Craniovertebral junction is affected by several disorders, which cause osseous expansion. Rickets (and osteomalacia) is one of them.2 Bony thickening and osteosclerosis can occur from abundant production of incompletely calcified bone matrix. This can result in foramen magnum stenosis, canal stenosis and basilar impression.2 Basilar impression secondary to osteomalacia has been reported in rickets. It may develop in the first two years of life when the softened skull base is unable to support the disproportionately heavy weight of infant’s head. It has been suggested that some of the instances of the primary basilar impression may indeed have been due to rickets, which has now been corrected. Chiari malformation also has been described.2

The stability of craniovertebral junction is dependent on the competence of the ligaments of the region. Atlanto-axial dislocations or atlanto-occipital dislocations may result from rickets. Atlanto-occipital dislocation is generally fatal. Patients surviving atlanto-occipital dislocation for the first 48 hours may have a good outcome.3 Upto 25% may survive neurologically intact and another 25% may survive with minor deficits. Our patient became symptomatic following a trivial fall and slowly improved on his own. We feel that it is likely to be a chronic dislocation and the fall may have precipitated acute decompensation. Traction for initial stabilization and correction followed by internal fixation should be considered for the patients with gross malalignment and neurological deficits.3 Traction was deferred in our patient because of danger of pin penetration (due to bone softening). The aim of surgery in this patient was to decompress the craniovertebral junction. In osteomalacia due to non-healed rickets where fixation and fusion of the graft are unlikely to be satisfactory, we recommend initial decompression and treatment of rickets. Once the rickets heals, the need for fusion may be considered if required.

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References