Case Report

Cranio-vertebral junction anomaly – presenting as cervical myelopathy

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Abstract

Antlanto-axial dislocation with blocked and ankylosed C2-4 vertebrae causing severe canal stenosis with compression of cervico-medullary junction has been reported. The knowledge of such anomaly may be importance for radiologists, anaesthesiologists, orthopaedicians and neurosurgeons because cranio-cervical junction anomaly may result in sudden unexpected death. It can also result in dysphagia, dysarthria due to compression of cranial nerves and quadriparesis due to compression of spinal cord. We here report a rare case of cranio-vertebral junction anomaly with blocked and ankylosed C2,3,4 vertebrae with evidence of atlanto-axial dislocation causing compression of cervico-medullary junction and leading to cervical myelopathy.

Keywords: Atlas, axis, cervico-medullary junction, atlanto-axial dislocation, cervical myelopathy.

Introduction

Cranio-vertebral junction is a collective term that refers to the occiput, atlas, axis and supporting ligaments. It encloses the medulla, spinal cord and lower cranial nerves. The occipital bone is composed of basioccipital, exooccipital and supraoccipital portions enclosing the foramen magnum. Several types of developmental deficiencies have been described including total aplasia, keller type aplasia with persistence of posterior tubercle aplasia with unilateral/bilateral remnant, midline rachischisis and hemiaplasia or partial aplasia of the posterior arch. Patients with cranio-vertebral joint anomalies exhibit the first neurological signs and symptoms usually no sooner than second decade. We here report a rare case of cranio-vertebral junction anomaly with blocked and ankylosed C2,3,4 vertebrae with evidence of atlanto-axial dislocation causing compression of cervico-medullary junction and leading to cervical myelopathy.

Case Report:

A forty five year old male was admitted in MMIMSR, Mullana on 27th July 2013 with chief complaint of weakness in all four limbs for last two years which was insidious in onset and gradually progressive. It was not associated with pain or loss of sensation. There was no history of trauma,
surgery, fall. No history of faecal or urinary incontinence. No history suggestive of any chronic illness like diabetes, tuberculosis, hypertension, bronchial asthma. Patient was reformed chronic smoker, non-alcoholic and non-vegetarian by diet. On admission, vitals were stable with BP – 110/70 mm of Hg and Pulse rate of 82/min. On examination, the patient was conscious and was well oriented to time, place and person. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy and pedal edema. JVP was not raised. Respiratory system examination revealed decreased air entry bilaterally with normal vesicular breath sounds. Cardio vascular system and per abdominal examination was within normal limits. Central nervous system examination revealed normal higher mental functions, normal sensory system and normal cranial nerves. Motor examination revealed hypertonia in all four limbs with power 5/5. All deep tendon reflexes were brisk with presence of ankle clonus. Superficial abdominal reflexes were absent. Planter reflexes were bilaterally extensor. Scissor gait was present. There were no signs of meningeal irritation. All routine investigations were within normal limits. X-ray cervical spine showed fusion of C2,3,4 vertebrae with reduced inter-vertebral disc spaces between C4 – C5 and C5 – C6 (Fig.1) MRI showed cranio-vertebral junction anomaly with blocked and ankylosed C2-4 vertebrae with evidence of atlanto-axial dislocation with dens and axis causing severe canal stenosis with compression of cervico-medullary junction and compressive myelopathy (Fig.2). Degenerative changes were noted in rest of the spine.

**Discussion:**

Cranio-vertebral abnormalities have been recorded in morphological and clinico-radiological studies for many years. There is increased prevalence of associated fusion of axis and third cervical vertebra in association with atlanto-occipital assimilation. The signs and symptoms of pyramidal tract, anterior bulbar tract and cranial nerve involvement may be present. In patients with anomalies with cranio-vertebral junction, the natural history is not understood clearly. Many patients are asymptomatic and remain so throughout their lives. Clinical manifestations like restricted neck movements, neck pain, headache, numbness, weakness in limbs, tinnitus, dysphagia, dysarthria develop only with advanced age or after trauma. Symptoms of clinical importance are caused by four principle mechanisms that are mechanical compression of nervous structure, mechanical compression of vascular structures especially of vertebral arteries, marked instability or in contrast elevated rigidity and disturbance of mechanical mobility of skull joints.
Conflict of Interest
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