



Quad Fever as a Rare Presentation of Os Odontoideum- A Case Report

Authors

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Abstract

Os odontoideum is a rare craniovertebral anomaly of the second cervical vertebra presenting with restricted neck movements, neck pain, brainstem dysfunction, cranial nerves involvement, even myelopathic weakness of limbs. Fever with quadriparesis is not one of its presentations. We are reporting a case of a 15yr old male patient who presented with fever and quadriparesis. He was diagnosed on MRI to have Os odontoideum. Fever in this case played the role of a red herring. During the course of hospital stay, he developed hypoventilation syndrome; he was placed on ventilatory support and subsequently succumbed to ventilator associated pneumonia.

Introduction

Cranio-vertebral anomalies (CVJ) are congenital or acquired abnormalities of the occipital bone, foramen magnum or first and second cervical vertebra that decrease the space for the lower brainstem and cervical cord. Os odontoideum is one of its rare types. We present first case of os odontoideum presenting with quad fever and quadriparesis.

Case Presentation

A 15year old boy presented with complaints of moderate to high grade fever that was unrecorded, not associated with chills, rigors, rash, weight loss, sore throat, burning micturation, seizures. He also complained of occipital pain and headaches. The occipital headache was moderate to severe in nature, dull-aching type, radiated to the upper back, persisted for the whole day and was only relieved on analgesic medication. There was no history of altered behaviour, seizures, diplopia, nausea, projectile vomiting. This was followed by

gradually progressive quadriparesis. The weakness was insidious in onset; started from the right upper limb, progressed to the right lower limb, and then involving the left lower limb and finally the left upper limb over a period of two months. At the beginning, the patient could go about his daily activities with some difficulty but soon even going to the toilet became a chore as his legs could not support his body weight. He has become bed ridden for the past two weeks and cannot move his legs sideways or turn in the bed by himself. Patient also complained of paresthesias below his neck involving his whole body. There is no history of any level of sensory loss, bowel/bladder involvement, root pains. The patient later complained of difficulty in breathing. There is no history of cough, sputum, chest pain, wheezing, edema, palpitations, orthopnea, paroxysmal nocturnal dyspnea, hemoptysis, smoking. On physical examination, he was a young boy of average built and nutrition; appeared dyspneic but not cyanosed. There was no pallor,

icterus, lymphadenopathy but he had a raised body temperature of 99.8°F. His hair line was below fourth cervical vertebra (figure A) and his body to neck length ratio was 15.6 suggesting a short neck. Neck rigidity was absent but neck movements were restricted. There was no deformity in his spine. His higher functions and cranial nerve examination was normal. There was no horner's syndrome or downbeat nystagmus. He had spastic hypertonia and asymmetric power loss



Figure A: Low hair line and short neck

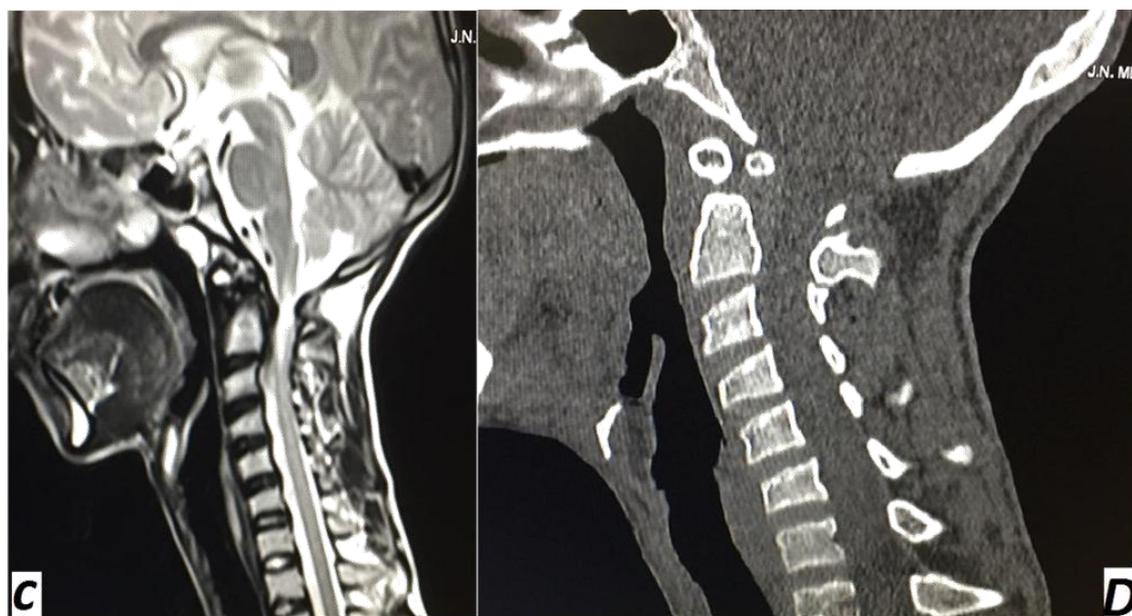
in all four limbs; deep tendon reflexes were brisk in all four limbs and superficial reflexes were absent; Babinski's reflex was positive and there was a loss of vibration upto the cervical vertebrae. Wasting of thenar and hypothenar muscles of both hands was present (figure B). The patient was unable to hold his breath for up to 20 seconds and he could not blow out a candle. The respiratory and cardiovascular systems were entirely normal.



Figure B: Wasted thenar and hypothenar muscles

The investigations include CBC Hb 11.2g/dl TLC 6,800/mcl DLC P69 L30 E01 ESR 12mm/hr; LFT ALT20IU/L AST17IU/L ALP7KAU/dL BIL1.2mg/dl; RFT BU28mg/dl S.CREAT 0.84; CSF Prot 20.4mg/dl Sugar 58.2mg/dl TLC 2 cells/mm³; Blood cultures, Urine cultures and CSF cultures were all sterile. MP smear, widal serology, dengue serology were all negative while CRP was normal. MRI brain was normal while that of the cervical spine revealed severe cervicomedullary junction compression with thinning and atrophy of the compressed segment (Figure C). CT was done to completely assess the associated bony abnormality. Plain CT neck revealed Os Odontoideum with rounding of

anterior arch of atlas (Figure D). Posterior ADI was reduced which is due to increased mobility of os odontoideum with ligamentous hypertrophy. As the patient went into hypoventilation due to respiratory muscle weakness, he was placed on ventilator support. No neurosurgical intervention could be done as the patient was unfit for surgery.



Sagittal T2W MRI (**Figure C**) CT image (**Figure D**) in a 15 year old patient with quadriplegia showing marked ligament thickening, spinal canal narrowing with cord compression and myelomalacic changes CT revealed rounded bony fragment lying above the base of dens. Dens is hypoplastic, smooth and well corticated and anterior arch is hypertrophied and rounded differentiating this condition from fracture.

Discussion

Os odontoideum is a rare CVJ anomaly of the 2nd cervical vertebra characterized by the separation of a portion of the odontoid process called dens from the body of the axis. The little bone can be identified by a small oval corticated ossicle whose size may vary. The transverse atlantal ligament becomes ineffective at restraining the atlanto-axial motion which may lead to compression of the cervical cord or vertebral arteries. The term was first coined by Giacomini in 1886⁽¹⁾. Os odontoideum may be asymptomatic and may be discovered incidentally on imaging⁽²⁾. Cervical pain was found to be the most common symptom of os odontoideum (64%) in a 2011 review of 78 patients. Sometimes it may be the only presentation^(1,3). The same review revealed headache as another symptom even though it was only in 2 patients⁽¹⁾. As headaches can be caused by many other innocuous conditions and have a high lifetime prevalence of 93-98% (of all types), they are far from being specific to os odontoideum⁽⁴⁾. A wide range of neurological deficits like transient quadriplegia, tetraplegia,

bulbar involvement and central cord syndrome may also result. Eighteen patients (23%) in the 2011 review presented with one of these neurological deficits while fifteen (19%) had a past history of one them^(1,5). Hereditary diseases such as Down syndrome, Klippel-Feil syndrome, Morquio's disease, multiple epiphyseal dysplasia, pseudoachondroplasia, achondroplasia, Larson syndrome, and chondrodystrophia calcificans also have os odontoideum as a clinical feature. Abnormally increased laxity of the ligament and incomplete ossification of the odontoid process is said to lead to Os odontoideum precipitated by trauma to the cervical spine⁽⁶⁾. Our patient presented with fever and quadriplegia. Keeping the Indian context in mind, the following differential diagnoses were made: 1.) Cervical epidural abscess at C2-C3 vertebra (?Tubercular) 2.) Pott's spine with collapse and compression or fracture and dislocation of C3/C4 vertebrae 3.) Meningomyelitis (?Tubercular/viral) 4.) CVJ anomaly. But neck rigidity, cough, sputum, weight loss, lymphadenopathy were all absent. CBC, ESR, CRP, chest x-ray and CSF examina-

tion were normal. X-ray cervical spine did not reveal any fracture or dislocation. The other findings were cervical pain, restricted neck movements, a low hairline, a short neck, quadriparesis and difficulty in breathing which supported the diagnosis of a CVJ anomaly. MRI cervical spine proved the existence of os odontoideum. There still remained no explanation of the fever as CVJ anomalies do not present with fever. The investigations failed to prove an inflammatory basis of the fever. It has been recorded in case reports that high cervical cord injuries are associated with fevers due to thermodyregulation caused by autonomic nervous involvement in the brainstem and upper cervical cord. Such fevers have been named “quad fever”^(7,8). This could be the non inflammatory and non-infectious cause of fever in this patient. Fever in this case had been the red herring which had led us to believe the lesion was infectious. Os dontoideum presenting with quad fever is a rare occurrence and has not been reported before.

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