

Basilar invagination in rheumatoid arthritis: A silent crusader

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A 42-year-old male was referred to the rheumatology clinic for inflammatory polyarthritis associated with multiple deformities for 10 years. His current complaints were the inability to perform overhead shoulder movements and neck pain for 2 years. The patient was a diagnosed case of seropositive rheumatoid arthritis and was on alternative medications, herbal, and Ayurvedic treatments. He had received conventional disease-modifying anti-rheumatic drugs intermittently; however, he was non-compliant with the same. He

complained of neck pain with stiffness and radiation of pain to the bilateral shoulders. He had difficulty in raising his shoulders overhead but had no weakness of lower limbs, sensory loss, or bowel and bladder incontinence.

On examination, he had severe restriction of bilateral shoulders and wrists (Fig. 1a-d). His Lhermitte's sign and Hoffman's sign were positive, suggesting cervical cord compression. His imaging of the cervical spine (Figs. 2 and 3) suggested upward migration of the odontoid process, causing basilar invagination (BI) and medullary compression.

Based on the imaging, he underwent posterior cervical spine decompression and fixation under neurosurgery. He was also restarted on tablets of methotrexate, hydroxychloroquine, and tofacitinib for adequate disease control.

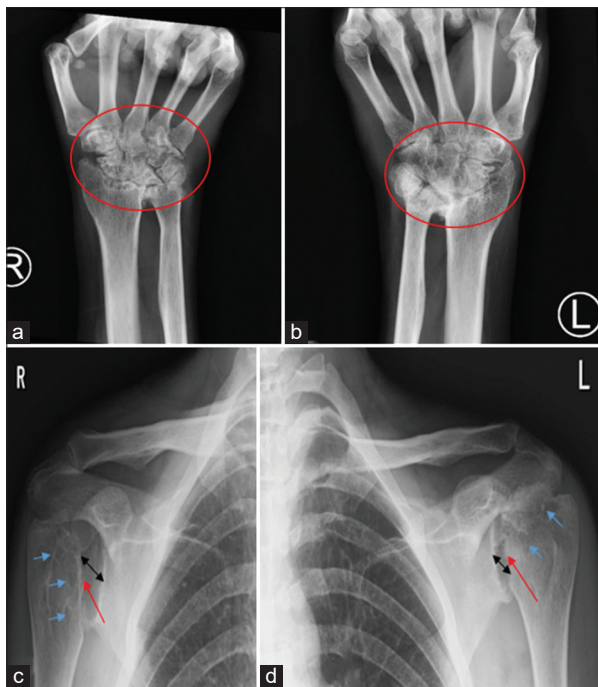


Figure 1: (a and b) Radiographs of bilateral wrist joints showing multiple erosions affecting radio-ulnar and carpal bones with joint space narrowing, sclerosis and ankyloses (red circles); (c and d) Radiographs of bilateral shoulder joints showing complete resorption of right humeral head (red arrow), multiple cysts (blue arrows) and widening of glenoid cavity (black double arrow head). The left shoulder shows resorption of the left humeral head (red arrow), cystic changes (blue arrows), and pseudo-widening of the glenoid cavity due to humeral head and acetabular erosions (black arrow heads). Deformity of the bilateral shoulder joints is noted as well

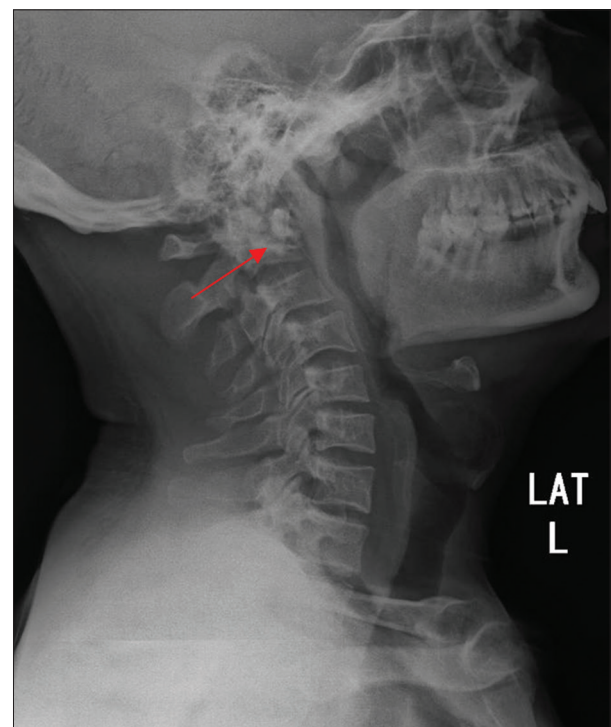


Figure 2: Lateral radiographic view of cervical spine showing upward migration of the odontoid process (red arrow), suggestive of basilar invagination

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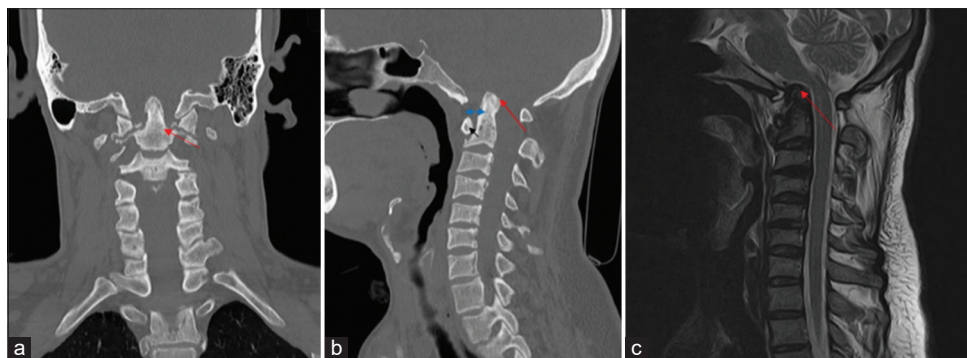


Figure 3: (a) Coronal computed tomography (CT) image of neck showing upward migration of odontoid process with basilar invagination (red arrow) “pencil tip abnormality;” (b) Sagittal image of CT neck showing upward migration of odontoid process (red arrow) crossing the Chamberlain line (black dotted line) for >3 mm, suggesting basilar invagination. The image also shows normal atlanto-axial distance (<3 mm, blue double-headed arrow) and presence of erosions over the C2 vertebral body (black arrow); (c) Corresponding midline sagittal image of magnetic resonance imaging cervical spine T2-weighted sequence showing upward migration of odontoid process compressing cervico-medullary junction (red arrow)

BI occurs due to cephalad migration of the cervical spine, causing narrowing of the foramen magnum. Most commonly, it occurs as a developmental anomaly or may develop later due to Chiari malformations [1]. It is a rare and late complication of untreated rheumatoid arthritis that occurs due to erosive changes, ligament and tendon instabilities, and cartilaginous destruction affecting the cervical spine, predominantly the atlanto-axial and occipito-cervical joints [2]. It can occur in up to 20% of cases of rheumatoid arthritis; however, early diagnosis and management can prevent this sequel [3]. Early symptoms like occipital headache, neck pain or stiffness, as in our case, can be subtle and missed. Progressive medullary compression can lead to myelopathy and cranial nerve palsies that require urgent neurosurgical intervention to decompress and stabilize the cervical spine [4].

Conventional radiography is not adequate to screen and diagnose BI [5]. Magnetic resonance imaging (MRI) is the gold standard to assess for odontoid pannus, cord compression, inflammatory changes, and overall spinal cord involvement. MRI also helps to delineate Chiari malformations and syringomyelia [1,5]. Apart from MRI, dynamic computed tomography can help to assess cranio-vertebral junction instability, bone instability, and the magnitude of bony destruction [1].

This case highlights importance of adequate disease control and patient compliance in rheumatoid arthritis to prevent delayed articular complications as shown in the images.

AUTHORS' CONTRIBUTORS

DM and RV were involved in the management of this patient. AS and DBL have reported the radiographs, CT, and MRI of this patient. All authors have reviewed the manuscript, images, and legends for publication.

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