

Uncommon Presentation of Ankylosing Spondylitis–Primary Stenosis at C1 Arch

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Aim: The object of this case report is to report primary C1 arch stenosis as a cause of myelopathy in an ankylosing spondylitis patient.

Background: Myelopathy at the level of the atlas in ankylosing spondylitis is commonly caused by atlantoaxial or occipitocervical subluxation, retro-odontoid pannus formation, cranial settling/basilar invagination, odontoid fracture. Myelopathic features are often neglected and are attributed to sacroiliac and lumbar spine involvement initially in these patients.

Case report: A 54 years old patient with long-standing ankylosing spondylitis presented with history neck pain with restriction of neck movements, gradually progressive weakness and stiffening of all four limbs. On examination, the patient had myelopathic features. Computed tomography (CT) of cervical spine showed straightening of cervical spine with severe stenosis at the C1 and bamboo stick appearance of a spine. The Magnetic resonance imaging (MRI) showed cord compression and T2 cord signal changes at the C1 level. The patient underwent C2 laminectomy and C1 posterior arch excision in neutral prone position. Postoperatively, the patient demonstrated improved limbs activity with a reduction in the flexor spasm.

Conclusion: Primary C1 arch stenosis may be one of the causes of myelopathy in ankylosing spondylitis patients and timely diagnosis and intervention benefits the patient and reduces morbidity.

Keywords: Ankylosing spondylitis, C1 arch stenosis, Enthesopathy.

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BACKGROUND

Inflammatory enthesopathy progressing to ossification and ankylosis is the pathologic basis for the Ankylosing

spondylitis (AS).¹ In the spine, the process of ankylosis usually begins in the lumbar spine and progress cranially to involve thoracic and finally, the cervical spine.²

We report a patient of ankylosing spondylitis presented with myelopathy due to primary C1 arch stenosis.

CASE REPORT

A 54 years old patient with long-standing ankylosing spondylitis was referred to our hospital. He was presented with history neck pain with restriction of neck movements for 10 years, gradually progressive weakness of all four limbs for 2 years. He was bed bound for 4 months. He had developed urinary bladder and anal sphincter dysfunction for 3 months.

On examination, active neck rotation and bending movements were not possible. Examination of cranial nerves was normal. The tone was increased in all four limbs and there were flexor spasms. Bilateral muscle power testing showed: deltoids grade 3/5, biceps 3/5, triceps 3/5, a hand grip 50%, iliopsoas muscle 4/5, knee and ankle 4/5. Sensations were reduced caudal to C4 dermatome. All deep tendon reflexes were brisk. Babinski's sign was present on both sides.

CT-scan (Fig. 1) revealed straightening of cervical spine with severe stenosis at the C1 and bamboo stick (completely fused spine) appearance of vertebral body secondary to ligamentum ossification (Fig. 1). C1 posterior arch was seen under C2 lamina. The MRI showed cord compression and T2 cord signal changes at the C1 level (Fig. 1).

The patient underwent C2 laminectomy and C1 posterior arch excision (Fig. 1) in neutral prone position. Intraoperatively C1 posterior arch was seen anterior to C2 posterior elements compressing the dura. After decompression dura expanded and was pulsatile. Histopathological evaluation of tissue around dura showed large areas of fibrosis with focal ossification involving bones. Granulation tissue was not seen, there by excluding active stage of disease. These features were consistent with chronic stage of Ankylosing spondylitis.

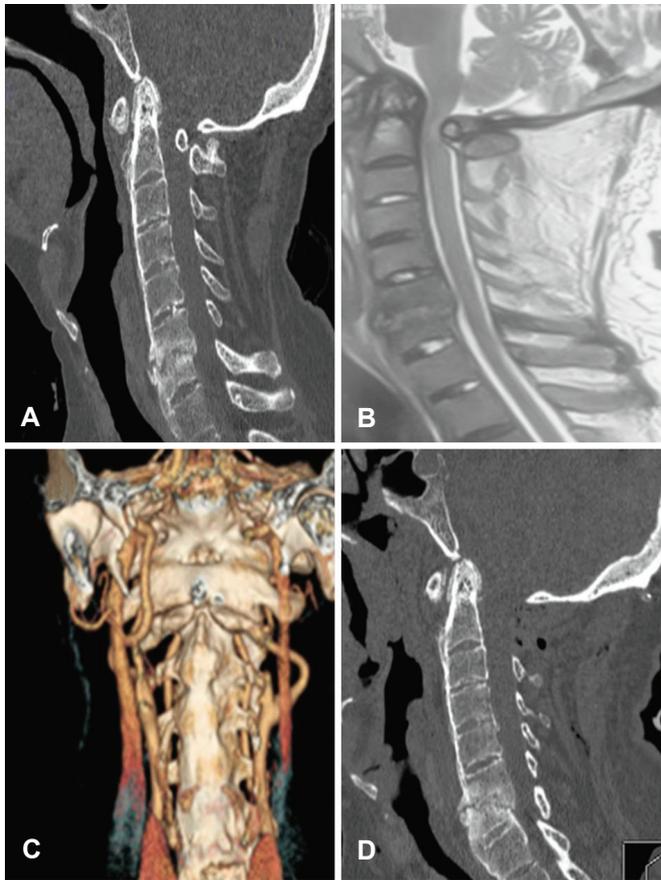
On follow-up at 1 year, the patient demonstrated improved limbs activity with a reduction in the flexor spasm.

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Figs 1A to D: In ankylosing spondylitis patient (A) Preoperative CT mid-sagittal cervical spine showing straightening and ankylosis of cervical vertebrae with canal stenosis at atlas; (B) MRI showing cord compression at atlas due to posterior C1 arch; (C) Surface shaded display coronal CT cervical spine showing fusion of posterior spinous processes; (D) Postoperative CT cervical spine showing absence of C1 posterior arch and C2 posterior elements.

DISCUSSION

High cervical stenosis is a rare entity, which can be developmental or acquired. Literature showed cervical myelopathy at the level of the atlas are caused by trauma, inflammatory diseases like rheumatoid arthritis, Paget's disease; neoplasm such as meningioma, bony tumors (aneurysmal bone cyst, osteoma) or metastasis; infective pathology such as tuberculosis.

Chronic inflammation of the entheses and new bone formation are two cardinal features of Ankylosing spondylitis. Cervical spine involvement in ankylosing spondylitis increases with age and disease duration.³ It is more common in patients with the symptomatic and severe forms of the disease. The craniovertebral junction is the most mobile region of the cervical spine, and enthesopathy involving it needs particular attention as it may restrict the mobility of joint.⁴ More importantly, enthesopathy at this level will cause high cord compression.

Review of literature showed myelopathy at this level in ankylosing spondylitis is caused by atlantoaxial or occipitocervical subluxation, retro-odontoid pannus

formation, cranial settling/basilar invagination, odontoid fracture.⁵ Isolated primary C1 arch stenosis as a cause of myelopathy in ankylosing spondylitis patients has not been reported till now. C1 stenosis with myelopathy due to C1-2 odontoid pannus causing anterolisthesis of C1 in a patient with ankylosing spondylitis is not an uncommon findings in AS. This is often due to the loss of mobility of cervical segments from C2-T1 which leave C1-2 articulation as the only mobile segment and thus subject to higher risk of arthritic degenerative changes. This patient had not shown any evidence of anterolisthesis on dynamic X-ray. This patient had a canal diameter of 0.5 cm with normal atlanto-axial distance.

In this patient, myelopathic features were initially neglected and were attributed to the sacroilitis and lumbar spine involvement as they are more common in these patients. Only after the patient developed flexor spasms, he was evaluated with MRI cervical spine and diagnosed to have C1 arch stenosis. Atlantodental ossification in long-standing disease may also contribute to the myelopathy in pre-existing narrowed canal due to C1 arch stenosis. Following posterior decompression, patient showed a reduction in spasticity and improvement in limbs activity.

The atlanto-occipital joints are the most frequently affected component of craniovertebral junction in AS patients.⁶ In severe disease, the involvement of the transverse ligament results in reduced elasticity and increased fragility of it. This may lead to ligament rupture and atlanto-dental dislocation. AS patients are also more prone for fracture as the spine is rigid and increased incidence of osteoporosis.⁷ This also requires special consideration during careful positioning in operating room.

CONCLUSION

Isolated C1 arch stenosis may be one of the causes of myelopathy in ankylosing spondylitis and decompression at C1 will reduce the morbidity in these patients.

CLINICAL SIGNIFICANCE

Clinicians need to be vigilant to diagnose myelopathy early as its presentation is often initially neglected and is attributed to the sacroilitis. Though myelopathy is commonly caused by inflammatory enthesiopathy, possibility primary C1 arch stenosis needs to be consider as differetial.

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