Unusual presentation of more common disease/injury

Odontoid pannus formation in a patient with ankylosing spondylitis causing atlanto-axial instability

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Ankylosing spondylitis is one of the commonest inflammatory diseases of the axial skeleton and can be complicated by atlanto-axial instability. This serious and likely underestimated complication can be easily overlooked. However, there are clear features which can help alert suspicion to initiate the appropriate investigations with imaging that is very effective at diagnosing and assessing this complication. The authors report an unusual case where odontoid pannus formation, akin to that seen in rheumatoid arthritis, was the underlying cause.

BACKGROUND

We present a case of atlanto-axial instability as a result of cervical pannus formation in a patient with ankylosing spondylitis (AS).

We decided to write up this case and explore the relevant literature in order to increase the awareness of a serious and likely underdiagnosed complication of a common disease; it can be easily overlooked as most patients with AS experience neck pain. However, there are clear features which can help alert suspicion and subsequent investigation with imaging that is very effective at diagnosing this complication. A further reason for submitting this case was that pannus formation of the odontoid in AS, as that seen in rheumatoid arthritis, is not well reported and was considered the cause of our patient's atlanto-axial subluxation.

CASE PRESENTATION

The patient was initially diagnosed with AS in 1996 when he was 18-years-old. Pelvic x-ray demonstrated bilateral sacroilitis. He was HLA B27 positive. There was no peripheral joint or ocular involvement but he had had episodes of plantar fasciitis and costochondritis. He had no other systemic illnesses. He was treated with diclofenac and began a rehabilitation programme which he continued to attend intermittently over the next decade.

In 2007, he was referred to our department with ongoing back pain and stiffness, having foregone several clinic appointments in other regional services. He had moderate to severe restricted lumbar spine movements. His bath ankylosing spondylitis disease activity index (BASDAI) was 5.09 and spinal pain score 6.8. Radiography showed grade 3 sacroilitis on the left and grade 4 on the right. There was no

Figure 1 (A, B) Lateral view plain x-ray of C-spine showing increased anterior atlanto-odontoid distance of approximately 8 mm.

evidence of spondylitis on whole spine imaging. He continued physiotherapy and was commenced on meloxicam.

In September 2010, he started experiencing severe, intractable neck pain radiating to the occiputs and down the cervical(c) spine. He required morphine for effective analgesia. His BASDAI was 6.26 and spinal pain 9.3. Treatment with anti-TNF was considered and a c-spine x-ray was ordered. Neurologically there were no deficits educed.

INVESTIGATIONS

The x-ray revealed atlantoaxial subluxation (AAS) of 8 mm (figure 1A,B). Subsequent MRI scan (figure 2A,B) confirmed AAS with marked intraspinal pannus formation and synovitis around the odontoid peg without neurological compression. C-spine CT scan (figure 3) did not show erosions

of the odontoid. Dynamic C-spine x-rays (figure 4) showed an anterior atlanto-odontoid distance (AAOD) of 1.1 cm on flexion and 0.65 cm on extension, consistent with AA instability.

OUTCOME AND FOLLOW-UP

He was fitted with a firm collar and a spinal surgical opinion was sought. Surgery was delayed initially for 3 months as his symptoms improved. However, by December 2010, with a progression of his symptoms, surgery was felt to be imperative and he underwent posterior spinal fusion with an uncomplicated postoperative recovery (figure 5). Treatment with α tumour necrosis factor (α TNF) was delayed until postoperative spinal bone fusion was achieved.

Figure 2 A (Saggital) and B (Axial) show a T2-weighted MRI image of C-spine confirming atlanto-axial subluxation with marked pannus formation and synovitis around the odontoid peg. There was no spinal cord compression or evidence of odontoid erosions.

Figure 3 Saggital CT scan of C-spine not showing any evidence of odontoid erosions.

Figure 4 Dynamic plain x-rays of the C-spine showing an atlanto-odontoid distance of 1.1 cm on flexion and 0.65 cm on extension, consistent with atlanto-axial instability.

DISCUSSION

Though AAS is an uncommon but recognised complication of AS, seldom is it as a result of pannus formation akin to that seen in rheumatoid arthritis. Several reports have demonstrated AAS as the presenting feature of early AS,¹ though generally it is patients who are older and with longer standing disease that are affected. Anterior AAS is the most common form of subluxation; vertical subluxation is rare.² Ramos-Remus found the prevalence of AAS in AS was approximately 20% and was associated with advanced sacroiliitis and ossification of the posterior longitudinal ligament.³ In the 2 year follow-up, AAS progressed in the majority of patients who did not undergo spinal surgery.⁴

The suspected mechanism of subluxation and instability is localised synovitis producing laxity or rupture of the transverse ligament. Pannus formation, best shown on MRI, is rarely seen in AS but if present can cause myelopathy through direct anterior impingement. Singh describes a patient with AS who developed severe neck pain secondary to pannus disease around the dens without odontoid instability or subluxation; he responded well to immunosuppression alone.⁵

Intractable neck pain radiating superiorly towards the occiput is one of the earliest and most common symptoms of AAS (70% of patients).⁶ Neck pain can also be a symptom of active AS and thereby difficult to distinguish from AAS; the clue is in the intractable nature of AAS. Other symptoms that may help in distinguishing between the primary disease and secondary complication includes radicular pain, Lhermitte's sign, loss of fine motor control or gait changes, though these features are uncommon in the early stages of AAS. Clinical examination should be focused on

eliciting signs of myelopathy and associated complications caused by localised compression (eg, cranial nerve palsies, horner's syndrome); one report of an patient with AS with vertical AAS had bilateral hypoglossal nerve palsy.⁷

Radiological assessments of the degree of subluxation and presence of myelopathy, in addition to a thorough neurological examination, are crucial to establish the need for spinal surgery. The correlation between the degree of subluxation and neurological sequelae is variably reported. Generally an AAOD of >9 mm suggests severe instability prompting the need for spinal surgery. Some suggest that the posterior atlanto-odontoid distance (PAOD) correlates better with neurological symptoms, as it takes into account, as part of its measurement, the diameter of the spinal cord. A minimum PAOD of 14 mm is required to avoid cord compression. Boden assessed the predictive value of PAOD⁸; PAOD of <14 mm on plain x-ray had 97% ability to detect patients with neurological deficit. Furthermore, neurological recovery from surgery was unlikely if the PAOD fell below 10 mm and complete motor recovery occurred if the surgery was performed while the PAOD was >14 mm.

Treatment objectives are to decompress the neural elements and stabilise the spine to prevent further spinal cord injury. Though there are reports of pannus debulking using immunosuppression, only the former treatment objective (spinal decompression) can be achieved but not the latter (stabilisation). The subsequent AA instability due to transverse ligament weakness can only be corrected surgically.

Any AS patient with obstinate neck pain should alert the suspicion of AAS. Pannus disease is a rare feature in patients with AS. Close monitoring of those who do not have surgery is recommended as AAS is likely to progress.

Figure 5 Lateral view plain x-ray of C-spine after posterior spinal fusion surgery.

Learning points

- ▶ Odontoid pannus formation, as that seen in rheumatoid arthritis, can occur in AS.
- ▶ Atlanto-axial subluxation in AS is likely more prevalent than currently appreciated leading to an underestimation of a serious complication.
- ▶ Intractable neck pain should alert suspicion and prompt appropriate assessment.
- ▶ Though there are anecdotal reports of treating AAS with increased immunosuppression, surgery is still the treatment of choice to achieve spinal stability.

Competing interests None.

Patient consent Obtained.

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