Unusual presentation of more common disease/injury

An unusual cause of recurrent falls in an older gentleman

Alexander Brent,¹ Ruth Hartley²¹ FY1, Department of Elderly Care, Mayday University Hospital, London, UK² ST3, Department of Elderly Care, Mayday University Hospital, London, UKCorrespondence to Alexander Brent, ajb101@doctors.org.uk**Summary**

The authors present the case of an 82-year-old gentleman who was admitted through Accident & Emergency with a history of increasingly recurrent falls. A thorough history and examination led to the provisional diagnosis of cervical cord compression. Subsequent investigations confirmed an erosive abnormality around the odontoid peg, with an adjacent soft tissue mass. The patient underwent a partial odontoidectomy and the histological examination of the pannus revealed a basophilic crystalline arthropathy, consistent with pseudogout.

BACKGROUND

The occurrence of pseudogout in the craniocervical junction is extremely rare, with only 12 cases being reported before 2000.¹ Since 2000, only singular case studies have been reported, with approximately one a year worldwide.

Pseudogout typically presents as pain and swelling of acute synovitis, caused by crystal shedding into the joint.² There are no previous case reports in the literature of pseudogout presenting as recurring falls.

CASE PRESENTATION

An 82-year-old gentleman, previously fully mobile and independent, presented to hospital with increasing postural instability and falls, worsening in number and severity over the preceding 4 weeks. This was on the background of a 2-year history of increasing leg and arm weakness. On further questioning, the patient had also developed bilateral arm paraesthesia over the last 2 weeks but had not suffered from any neck or back pain. There was no prior injury.

On examination, the patient had bilateral limb weakness, hyperreflexia and upgoing plantars. This pattern was found in both upper and lower limbs but generally was more severe in the arms. Sensation was grossly intact although there was loss of vibration sense distally. These symptoms had not been reported by the patient until directly questioned at the time of the final fall.

INVESTIGATIONS

The patient had a full set of spinal x-rays. The lateral views demonstrated fusion of the bodies and posterior elements of C3 and C4, suspected to be a developmental abnormality, and severe degenerative changes of the lower vertebrae, leading to a provisional diagnosis of cervical spondylosis causing cord compression.

This diagnosis was supported by the CT images, which showed fused posterior elements of C2–C5, narrowing of C5/C6 foramina, osteoarthritis of the odontoid peg and generalised spondylotic changes. However, in light of the clinical picture and upgoing plantars, an MRI was requested.

Apart from the spondylitic features, the MRI also showed an irregular, eroded odontoid peg (figure 1). There was

evidence of a low-density, soft tissue pannus making contact with the anterior cord as it emerged from the skull base, leading to significant canal narrowing at the C1 level with effacement and signal change in the intrinsic cord.

The patient was referred to a specialist neurosurgical unit for urgent intervention. He underwent a partial odontoidectomy with removal of the posterior pannus. The histology report of the resected tissue showed focal necrosis with infiltration with a basophilic granular material. This comprised of birefringent crystals with rectangular and rhomboidal shapes, consistent with calcium pyrophosphate, which causes pseudogout.

DIFFERENTIAL DIAGNOSIS

The most common causes of cervical myelopathy include spondylosis, disc prolapse and trauma,³ with rarer causes including neoplasms and infection.² Pseudogout is defined as the deposition of calcium pyrophosphate dehydrate within the cartilage with the subsequent articular inflammatory reaction. It most commonly occurs in the knee or wrist of older women.² Symptoms are typically pain and swelling of acute synovitis caused by crystal shedding into the joint. As described, the occurrence of pseudogout in the craniocervical junction is extremely rare, and no previously reported cases have presented as recurrent falls.

In this case, it is important to differentiate whether the patient's symptoms were caused primarily by the spondyloarthropathy of the cervical spine or from the odontoid mass, especially considering the changes reported in the spinal images. The degree of onset of the patient's symptoms was too rapid and too constant to be consistent with that of spondyloarthropathy. One would expect degenerative changes like this to present over a longer period of time, causing intermittent symptoms, often linked to movement. Another distinguishing feature is that the operation was a partial odontoidectomy, where only the mass was removed and did not involve removal of syndesmophytes or osteophytes. Symptoms immediately improved from here on, albeit over a long period of time. Taking into account these factors and the extensive degree of calcium pyrophosphate crystals in the histological specimen, it was felt by the teams involved that this was a diagnosis of pseudogout. It is

Figure 1 MRI demonstrating soft tissue pannus around C2 causing deformity.

important to consider in this case that given the degree of spondyloarthropathy, it is actually very likely to have caused the formation of the calcium pyrophosphate crystals in the first place. This is a known and documented cause of pseudogout and thus the diagnosis would be that of secondary pseudogout.

Another important differential diagnosis to rule out in this case is that of an infective process causing an inflammatory mass. Clinically, the patient had no signs of osteomyelitis or septic arthritis. His inflammatory markers were not significantly raised and he showed no signs of a systemic inflammatory response. The pathological specimen sent to the laboratory had no bacterium present.

TREATMENT

The patient in this case benefited from neurosurgical intervention combined with subsequent rehabilitative care. Once the patient had benefited from all the occupational therapy and physiotherapy the hospital could offer, he was

transferred to a neurosurgical rehabilitation centre for specialist care.

OUTCOME AND FOLLOW-UP

The patient made significant improvements in power and function, but due to the duration of his symptoms originally he was still requiring intensive physiotherapy 4 months later to help regain mobility. He eventually managed to gain enough power and mobility to be moved to a local nursing home.

DISCUSSION

There are two clinically distinct presentations of pseudogout of the odontoid peg previously described. The more common of the two is the more fulminant presentation of crowned dens syndrome (CDS). CDS results in intermittent severe neck pain, fever and raised inflammatory markers, which mimics other diagnoses such as meningitis and giant cell arteritis.⁴ Much rarer is the presentation seen here,

where subtle signs of cervical spinal cord compression develop over years.^{5 6}

Learning points

- ▶ A detailed neurological history and examination is important in patients presenting with a fall.
- ▶ Eliciting an accurate history and thorough examination can be challenging in the older age group.
- ▶ Crystalline arthropathy can occur in any joint.

Competing interests None.

Patient consent Obtained.

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