Case Report

Diffuse Idiopathic Skeletal Hyperostosis with Cervical Spinal Cord Injury – A Report of 3 Cases and a Literature Review

S Sreedharan,[1] *MBBS*, YH Li,[2] *FRCS (Glas, Edin), M Med (Surg), FAMS*

Abstract

Introduction: Diffuse idiopathic skeletal hyperostosis (DISH), though common, is often asymptomatic. However, spinal hyperostosis can predispose the affected to chronic myelopathic symptoms and acute spinal cord injury. Clinical Picture: We report on 3 patients with DISH, who sustained traumatic cervical cord injuries. Two were tetraplegic at presentation. The radiologic findings of the patients are also discussed. Treatment: Both the tetraplegic patients were treated non-surgically in view of high surgical risk. Outcome: Both the tetraplegic patients died due to mechanical respiratory failure. Conclusions: The potential catastrophic neurological sequelae of DISH from relatively minor trauma must be understood. Further studies are needed to aid in evidence-based clinical management of asymptomatic patients with DISH.

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Key words: Forestier disease, Ossification, Posterior longitudinal ligament, Vertebral ankylosing hyperostosis

Introduction

Despite being a largely asymptomatic clinical condition, diffuse idiopathic skeletal hyperostosis (DISH) has been widely described in literature due to its often-interesting clinical manifestations. Here we describe 3 case reports of spinal cord injuries in patients with DISH who presented to Tan Tock Seng Hospital, Singapore, between March 2000 and April 2004. We also include a review of the diagnosis, epidemiology and common mechanisms of cord injury in DISH.

Case Reports

Case 1

Mr H, a 76-year-old Chinese gentleman, had a history of bilateral osteoarthritis of the knees and was on follow-up with a cardiologist for arrhythmia. He presented with a hyperextension injury to the neck, sustained when he tripped and fell forwards, landing on his left temple. He complained of transient weakness of all 4 limbs, with the upper limbs being weaker than the lower limbs, and was unable to walk immediately after the injury.

On presentation at the emergency department, Mr H was alert and communicative. There was a laceration on the left side of the forehead and the posterior aspect of his neck was tender. Neurological examination, however, was completely normal by the time of presentation. Power was full in all 4 limbs, proximally and distally. There was no sensory deficit. Anal tone was good with absence of saddle anaesthesia. Cervical spine radiograph (Fig. 1) showed a fracture through the bridging osteophyte between C3 and C4, with underlying ankylosing hyperostosis from the level of the first to the seventh cervical vertebra.

In view of a history of neurological loss, a magnetic resonance imaging (MRI) scan of the cervical spine (Fig. 2) was performed. This showed a fracture of bridging osteophytes at the C3-C4 level anteriorly with surrounding pre-vertebral oedema. There was, in addition, posterior disc protrusion and hypertrophy of facet joints at the same level, contributing to significant cord compression.

Clinically, however, from admission onwards, Mr H did not display any neurological deficits. The cervical spine was stabilised with an Aspen collar and ambulatory physiotherapy was started within 2 days. On outpatient follow-up after discharge, Mr H remained well. He was

[1] Department of General Surgery
Singapore General Hospital, Singapore
[2] Department of Orthopaedic Surgery
Tan Tock Seng Hospital, Singapore
Address for Reprints: Dr S Sreedharan, Department of General Surgery, Singapore General Hospital, Outram Road, Singapore 169608.
Email: sreedharan.sechachalam@singhealth.com.sg

offered decompression surgery for the residual spinal canal stenosis at the C3-C4 level but Mr H declined surgical intervention.

Case 2

Mr W, an 84-year-old Chinese gentleman, had underlying diabetes mellitus (DM), hypertension and ischaemic heart disease for which a coronary artery bypass grafting had been previously done. He presented with a cervical hyperflexion injury, sustained when he fell backwards off a bench, landing on his occiput. Mr W presented to the ED with numbness and weakness of all 4 limbs.

On examination, Glasgow Coma Scale (GCS) was 15. Neurological examination revealed power to be 0 throughout both lower limbs and in the left upper limb; power was 1 to 2 in right upper limb at the C4-C6 myotomes. Sensation to fine touch and pin prick was lost at all dermatomes below C3 to C4 bilaterally. Anal tone was lax on presentation.

Cervical spine radiograph (Fig. 3) showed a C5-C6 facet joint fracture and dislocation of 50%, and underlying ankylosing hyperostosis from C1 to C6. MRI (Fig. 4) showed an acute fracture through C6 involving the anterior-superior corner, extending into the C5-C6 intervertebral disc and running posteriorly; the fracture was associated with a facet joint dislocation. The spinal cord was compressed at the C5-C6 level with cord oedema present. The patient was started on intravenous methylprednisolone to reduce cord oedema. The C5/C6 fracture dislocation was reduced with skull traction, with sequential load increments by 5 pounds.

Due to his significant medical comorbidities, Mr W was considered a poor surgical candidate. This was explained to the patient and his family, who then opted for non-operative management. Despite the reduction in the fracture-dislocation, Mr W's neurological state declined: he became progressively less responsive and his GCS dropped from 15 to 3 in the first 2 days of admission. This was attributed to carbon dioxide narcosis secondary to mechanical respiratory failure. He was intubated and mechanically ventilated from the third day of admission. However, his condition deteriorated further and he passed away 2 weeks after admission.

Case 3

Mr P was a 69-year-old Chinese gentleman with hypertension and a history of stroke. He presented to the ED with complete paralysis after sustaining a hyperextension injury to the neck when he tripped and fell forward onto his forehead. At the ED, GCS was 15. Neurological examination revealed power of grade 0 in all 4 limbs. Sensation was absent bilaterally from the level of the C4 dermatome downwards. Anal tone was also absent. Cervical spine X-ray showed a fracture through the anterior column of hyperostosis at the level of the C3/C4 vertebrae and underlying DISH from the C1 to C7 vertebrae (Fig. 5).

MRI (Fig. 6) showed a focal area of spinal canal stenosis at C3/C4 due to both anterior and posterior compression by osteophytes and a thickened ligamentum flavum. There was cord compression and oedema of the cord at the same level. As with Mr W, Mr P's family opted for non-operative management in view of high surgical risk due to underlying medical comorbidities. Medical treatment of cord compression was initiated with intravenous methylprednisolone.

However, on the same day of admission, he developed mechanical respiratory failure. Again, this was likely secondary to involvement of the nerve roots for C3-C5. Mr P was mechanically ventilated for 3 months, during which time he succumbed to complications of deep venous thrombosis and multiple episodes of pneumonia, and eventually passed away.

Discussion

Other names for diffuse idiopathic skeletal hyperostosis (DISH) include Forestier's disease[1] and ankylosing hyperostosis. It is a well-defined syndrome with axial and peripheral manifestations of hyperostosis – these include hyperostosis frontalis, calcaneal spurs, post-hip surgery heterotopic ossification and anterior/posterior longitudinal ligament ossification.

The prevalence of DISH in the general population varies greatly depending on the location of the study. Prevalence rates of up to 27.3% in males and 12.8% in females have been quoted but a consistent finding is that males are more commonly affected – up to twice as often as females.[2,3]

DISH is uncommon in those below the age of 50 years, after which incidence increases with age. Incidence also increases with weight[4] in both sexes. In fact, the oldest described risk factor for DISH is obesity, which is mentioned in the first description of the syndrome.[1] In a case control study involving 131 subjects, the body mass index was significantly higher in those with DISH (27.8 versus 26.0).[4]

DM is commonly quoted as a risk factor. However, Vezyroglou et al[5] showed that while diabetics who are also hyperuricaemic and/or hyperlipidaemic have a significantly higher incidence of ankylosing hyperostosis, DM alone is not a statistically significant risk factor. Animal models have shown that high serum vitamin A levels increase the risk of DISH;[6] no such studies on humans were found in the literature. A recent review article of Forestier's Disease by Sarzi-Puttini and Atzeni[7] reiterated the greater prevalence of high body mass index, DM and hyperuricaemia in patients with DISH.

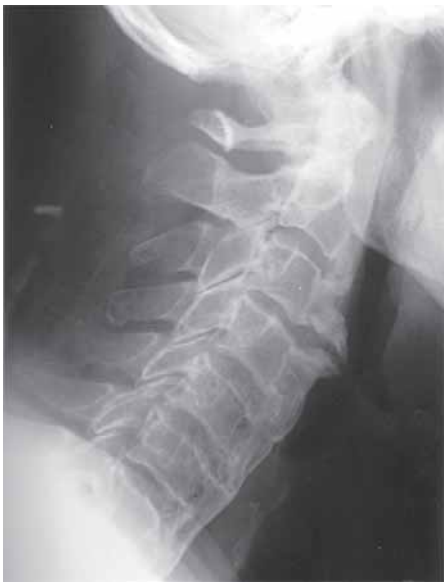

Fig. 1. Cervical spine radiograph of case 1.

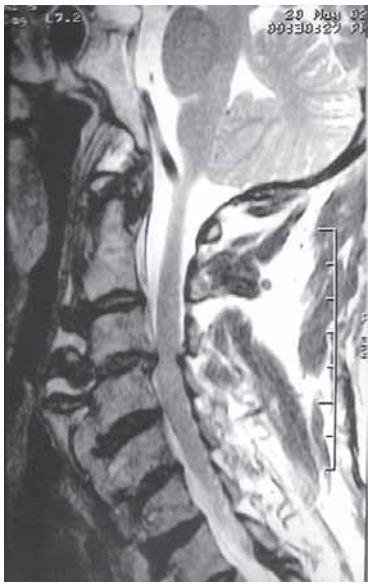

Fig. 2. MRI cervical spine of case 1.

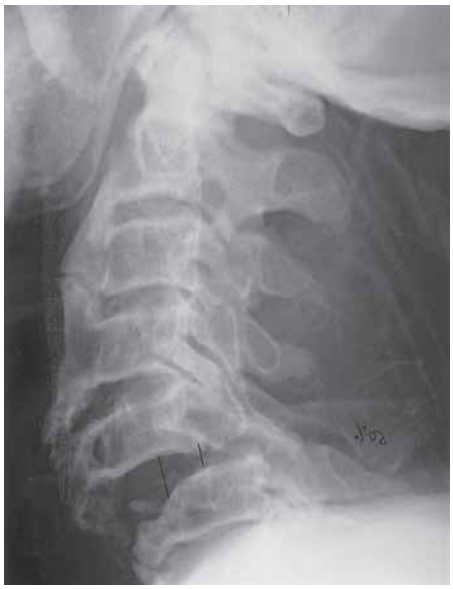

Fig. 3. Cervical spine radiograph of case 2.

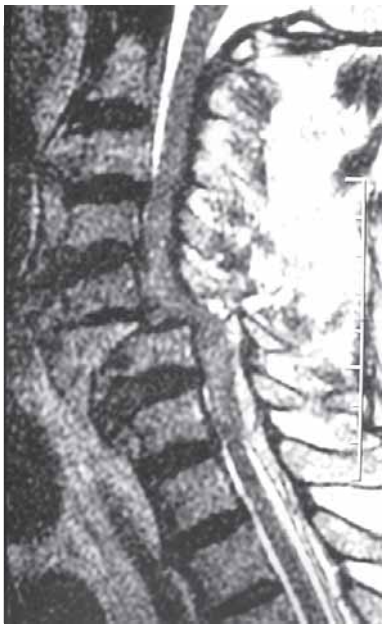

Fig. 4. MRI cervical spine of case 2.

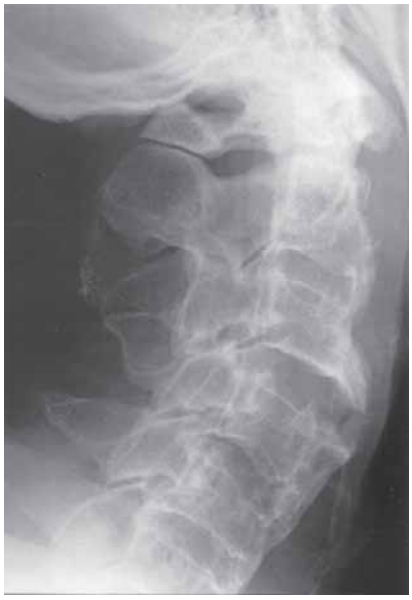

Fig. 5. Cervical spine radiograph of case 3.

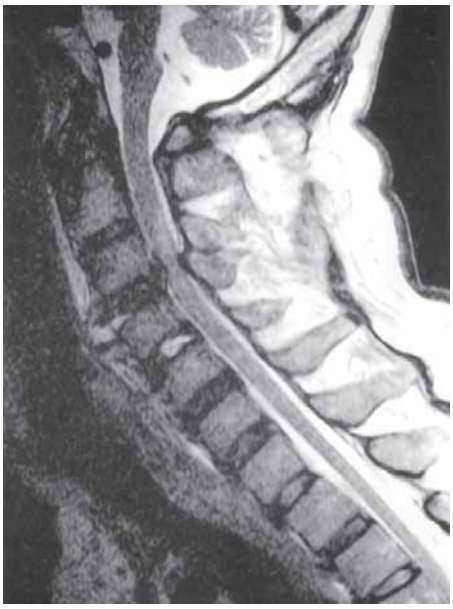

Fig. 6. MRI cervical spine of case 3.

The diagnosis of DISH is based on the following radiologic features:[8]

1. *New bone formation bridging at least 4 contiguous vertebral bodies (Resnick's criteria)*

   Thoracic spine is often an ideal region to display the bridging as it is relatively non-mobile, thereby allowing unhindered ossification from one vertebra to another. Appropriate X-ray views include antero-posterior and lateral views of the region of interest.[9]

2. *Absence of degenerative disc disease*

   This is imperative in distinguishing osteophyte formation from ossification of the spinal ligaments.

3. *Absence of inflammatory changes in facet or sacroiliac joints*

   This criterion differentiates DISH from its closest radiological twin, ankylosing spondylitis. Hence, if there is clinical suspicion of ankylosing spondylitis or vertebral X-ray is non-confirmatory for DISH, a pelvic X-ray will be necessary to exclude sacroiliitis. For this, an anteroposterior view of the pelvis will suffice – Battistone et al[10] claim detailed oblique projections do not provide additional benefit, as previously thought.

The majority of people with DISH are asymptomatic. Of those with clinical manifestations, neurological symptoms are the most common. However, subjects do occasionally present with other symptoms, including mechanical dysphagia, dyspnoea, stridor, thoracic outlet syndrome and heterotopic ossification after total hip arthroplasty.[11,12]

Neurological manifestations secondary to DISH are commonly due to involvement of the cervical spine, which is affected in 75% of patients with DISH.[13] These stem from:

1. Reduced flexibility of the spine, as in ankylosing spondylitis,[14] thereby allowing a trivial trauma to result in a fracture through the ossified ligaments. This acutely compromises the size of the spinal canal. Fractures of such nature in the cervical spine are associated with a higher morbidity and mortality than fractures through the normal spine.

   All 3 patients mentioned above suffered injuries of this nature. Mr W and Mr P developed mechanical respiratory failure likely due to ascending cord oedema involving the C3/C4/C5 levels (roots supplying the phrenic nerve) following the fractures.

2. Spinal canal narrowing secondary to ossification of anterior and posterior longitudinal ligaments. Patients present with myelopathy as a result. Furthermore, there is less potential space available within the canal such that cord compromise occurs sooner in the event of fractures and cord oedema (as in the 3 case studies above).

3. Atlantoaxial subluxation (AAS) of the cervical spine may rarely occur in subjects with DISH. The literature search yielded 2 case reports. Oostveen et al[15] described an 80-year-old lady with cervical radiculopathy. Chiba et al[16] described another patient with progressive myelopathy – the authors hypothesised that AAS is caused by the concentration of cervical motion from the atlanto-occipital to the C1-C2 joints as a result of the loss of cervical mobility below C2 (secondary to DISH). Narrowing of the spinal canal due to ossification of the spinal ligaments probably contributes to the neurological effects of atlantoaxial subluxation.

DISH is often an incidental diagnosis made from a chest radiograph taken for a non-related reason or a finding made at autopsy. Unless symptomatic, it does not require surgical treatment; for instance, decompression surgery may be offered for those with myelopathic symptoms. However, it is not altogether benign, as the case reports above and other previously published papers suggest.[17] Sharma et al[18] showed in a retrospective analysis that 15% of patients with DISH presenting to a particular neurosurgical unit had serious neurological manifestations requiring neurosurgical intervention.

Age may be a possible confounder in the relationship between DISH and the increased incidence of spinal cord injuries. While the incidence of DISH increases with age, so do other factors predisposing to increased risk of falls, such as failing eyesight and mobility, which could contribute to a higher incidence of traumatic spinal cord injury. DM is another known risk factor for DISH. Complications such as peripheral neuropathy, autonomic neuropathy, retinopathy, cataracts and hypoglycaemic episodes are a few of the reasons diabetics may be prone to falls.

As with increasing age, DM may then be an added risk for traumatic spinal cord injury in patients with DISH. By extension, hyperlipidaemia may be an additional risk factor for fall-related cord injury due to the link between hyperlipidaemia and stroke. A literature search did not yield studies on the presence or number of risk factors and their correlation with spinal cord injury in DISH. As such, although DISH is largely asymptomatic and usually diagnosed incidentally, it would be interesting to investigate if optimisation of the metabolic risk factors for DISH in patients with early diagnosis would reduce the rate of progression of hyperostosis and eventual likelihood of spinal cord injury.

The progression could be monitored using a grading scale, which may be of predictive value in determining a subject's risk of spinal cord compression. Possible points to consider in the scale include: number of bridging ossifications, degree of osteoporosis of the adjacent vertebral bodies and the diameter of the spinal canal. Those with higher risk could then be:

- educated on how to recognise myelopathic symptoms
- educated on fall prevention, and
- managed more aggressively for their coexisting medical morbidities, which may predispose them to falls, as mentioned above.

Conclusion

Diffuse idiopathic skeletal hyperostosis (DISH) is probably the commonest rheumatological disorder.[8] Although mostly asymptomatic, it can predispose the affected to catastrophic spinal cord injuries with risk of mortality, as seen in 2 of the 3 cases studied in this article. As such, patients should be informed about possible neurological sequelae from relatively minor trauma. We suggest possibilities for further studies correlating currently known risk factors for DISH and the role of their management in the prevention of progression and traumatic complications of this condition.

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