

An unusual case of stridor due to osteophytes of the cervical spine: (Forestier's disease)

K. PAPAPOSTAS, F.R.C.S., A. THAKAR, M.S., V. NANDAPALAN, F.R.C.S., G. O'SULLIVAN, F.R.C.S.

Abstract

Stridor is a noisy breathing caused by compromised airway in the larynx and trachea. The causes can either be due to intrinsic or extrinsic compression. Stridor resulting from extrinsic compression due to anterior cervical osteophytes is rare.

We report an unusual case of acute stridor due to an osteophytic mass in the cervical vertebrae resulting in a mechanical upper airway obstruction. The underlying pathology was Forestier's disease or diffuse idiopathic skeletal hyperostosis (DISH). Stridor is a rare manifestation of DISH and it certainly represents the most life-threatening one. Only a few cases have been reported in the English literature and are mainly secondary to impaired function of the vocal folds, or postcricoid ulceration and oedema. We present such a case, in that stridor was the result of direct airway obstruction by the osteophytic mass and an emergency tracheostomy had to be performed to establish an airway.

Key words: Stridor; Spinal Osteophytosis, Forestier's disease

Introduction

Forestier's disease (synonyms-ankylosing spondylosis; diffuse idiopathic skeletal hyperostosis (DISH)) is an idiopathic disorder, affecting mainly the middle-aged and elderly people, characterized by ossification along ligaments throughout the body, but most notably along the anterior longitudinal spinal ligament. It was first described by Forestier and Rotes-Querol in 1950. The prevalence rate of spine involvement on routine autopsy specimens was reported to be 12 per cent (Resnick and Niwayama, 1976). Most reported cases have had significant involvement of the cervical spine (Resnick and Niwayama, 1976; McCafferty *et al.*, 1995). Extraspinal calcification sites include the ligamentous attachments of the pelvis, calcaneus, tarsal bones, patella and olecranon (Resnick *et al.*, 1975).

The holding opinion regarding the pathogenesis of Forestier's disease, especially after the large cadaveric and radiological study by Resnick and Niwayama (1976), is that ossification in the cervical spine arises in the ligaments. Ossification of the anterior longitudinal ligament may progress to exuberant osteophyte formation thus causing compression of the pharynx and cervical oesophagus. In most cases symptoms are rarely significant enough to come to clinical attention.

Dysphagia is the commonest complaint (Barsamian *et al.*, 1985) and a number of case reports regarding this complaint have been documented in the literature under various surgical specialities (Meeks and Renshaw, 1973; Barsamian *et al.*, 1985; Brandenburg and Leibrock, 1986). However, stridor secondary to osteophyte compression has rarely been documented.

Case report

A 77-year-old man was referred from his General Practitioner to the Otolaryngology department at the Arrowe Park Hospital with an acute exacerbation of noisy breathing. The patient's past medical history included a three-year history of asthma and in 1993 he had been investigated for intermittent difficulty in swallowing. A barium swallow and meal performed at that time showed a large cricopharyngeal muscle impression and a small pharyngeal pouch, but no significant cervical spine abnormality. The patient had been managed conservatively because his symptoms were minimal. Examination of the patient revealed an inspiratory stridor with tachypnoea and tachycardia. There was no cyanosis. Fibre-optic endoscopy of the hypopharynx and larynx revealed that the posterior pharyngeal wall was bulging and abutting against the epiglottis with a small airway passage to the laryngeal inlet. Mucosa on the posterior pharyngeal wall appeared normal. The rest of the examination was normal. A lateral soft tissue X-ray of the neck demonstrated ossification of the anterior longitudinal ligament with multiple osteophytes and a prodigious osteophyte at the C3–C4 vertebrae causing compression and obstruction of the pharyngeal lumen (Figure 1). The radiographic appearance of the airway inferior to the epiglottis was normal.

Initially the patient was managed with 40 per cent humidified oxygen and intravenous dexamethasone, however, within the next two hours he developed increasing signs of fatigue. To relieve his airway obstruction a tracheostomy had to be performed and was carried out under general anaesthetic. Although the anaesthetist initially had difficulty in intubating, he managed to secure the airway with a microlaryngoscopy tube. A direct

From the Department of Otolaryngology – Head and Neck Surgery, Arrowe Park Hospital, Upton, Wirral, Merseyside, UK.
Accepted for publication: 30 October 1998.



FIG. 1

Lateral soft tissue neck X-ray demonstrating ossification of the anterior longitudinal ligament (C3–C7) and an osteophytic mass at C3–C4 leading to a posterior pharyngeal bulge causing airway obstruction. Note the normal appearances of the airway inferior to the epiglottis and the normal intervertebral disc spaces.

laryngoscopy performed at that time confirmed the narrowing of supralaryngeal airway due to extrinsic compression by anterior vertebral osteophytes. Distal to the bulge, the laryngeal inlet was normal with mobile vocal folds. Following the tracheostomy, the patient was able to breathe better and soon mastered the care of his stoma and tube change. With a phonatory valve tracheostomy tube he experienced little disability in communication. Dysphagia has not been a significant factor. In view of his age the options of further management of the cervical osteophytes were discussed with the patient and his relatives. The patient decided not to proceed with any surgical treatment for excision of the osteophytic mass. He was discharged home with the tracheostomy and on a recent review, he was coping well with his condition.

Discussion

The hallmark of Forestier's disease in the spine is the calcification of the anterior longitudinal ligament. Other characteristic radiological features are the preservation of the intervertebral disc height, the integrity of the apophyseal joints and the absence of outgrowths along the posterior aspects of the vertebral body. Extraspinal manifestations include calcification along the ligamentous attachments of the pelvis, calcaneus, tarsal bones, ulnar olecranon and the patella (Resnick *et al.*, 1975). Cervical

osteophytes are a common finding in the radiographs of the elderly patients. They may cause symptoms either by mechanical compression or by inducing an inflammatory reaction in the pharyngo-oesophageal wall (McCafferty *et al.*, 1995). However, the oesophagus being a very mobile structure, even large osteophytes may cause only minimal symptoms.

Dysphagia, mainly with solids, is the commonest symptom reported in the literature and is most likely to occur when the osteophytes are located in the regions where the oesophagus is tethered to the cervical spine, i.e. at the levels of the cricoid cartilage and the diaphragm (Deutsch *et al.*, 1985). Hilding and Tachdjian (1960) on reviewing 36 patients with dysphagia due to cervical osteophytes, found that 40 per cent of the patients had bony overgrowths at the C5–C6 vertebrae, 23 per cent at C4–C5, 14 per cent at C2–C3 and 14 per cent at C3–C4. Other less frequently presenting symptoms can be foreign body sensation and hoarseness. In 1990 Jonathan and Baer reported eight patients with a variety of complaints in whom the presence of significant cervical osteophytes were documented. The primary symptoms were foreign body sensation and dysphagia of varying degrees and one of the patients was described as mildly stridulous, with no further information given regarding his airway management. In the case we present the predominant involvement was at C3–C4, i.e. above the level of commencement of the oesophagus, and this may explain the reason dysphagia was not a symptom.

Although DISH is pathologically characterized by a very gradual overgrowth of osteophytes, it may occasionally present with acute symptoms. Upper airway obstruction due to this condition is rare. Hassard in 1984 reported two patients who developed airway obstruction and underwent emergency tracheostomy. Their radiographs showed prominent anterior cervical osteophytes. In the first patient, direct laryngoscopy showed a postcricoid ulceration and impaired vocal fold movement, presumably secondary to inflammation of the posterior crico-arytenoid muscle. The ulceration was believed to be caused by constant movement of the cricoid lamina over a projecting osteophytic mass. Treatment with antibiotics led to resolution of the ulcer. The patient then underwent surgical removal of the osteophytic mass. Vocal fold movement came back to normal and successful decannulation followed. Endoscopy of the second patient showed postcricoid ulceration and complete bilateral vocal fold paralysis. He was decannulated a few months later and panendoscopy showed a healed cricoid ulcer and improved vocal fold movement. Two further cases reported by another author (Verstraete *et al.*, 1998), manifested chronic dyspnoea secondary to bilateral vocal fold fixation at adduction.

Stridor caused by a large extraluminal osteophytic mass directly obstructing the supraglottic airway with no evidence of any associated vocal fold palsy, soft tissue swelling or ulceration of the pharyngeal wall has only rarely been reported in the English literature. We present such a case of this rare manifestation. Forestier's disease sometimes may be confused with ankylosing spondylitis which may be associated with similar clinical and radiological features. Forestier's disease is generally a disease of the elderly while ankylosing spondylitis affects young adults. As in ankylosing spondylitis, Forestier's disease does not cause sacroiliac joint ankylosis, facet joint ankylosis or a rise in the titre of acute phase reactants (McCafferty *et al.*, 1995). Another possible differential diagnosis is spondylosis deformans, in which the intervertebral disc spaces are also maintained but the anterior osteophytes are less prominent. The differences with

Forestier's disease are quantitative, the latter having more advanced radiological and pathological changes (Resnick and Niwayama, 1976). Hence, many clinicians nowadays do not consider spondylosis deformans a distinct clinical entity.

The presence of osteophytes secondary to degenerative joint disease or ankylosing vertebral hyperostosis is a common condition, mainly in the elderly. The majority of these patients are asymptomatic. Whenever an older patient presents with significant cervical or lumbar loss of mobility in the absence of a history of pain, the possibility of DISH should be considered (Rotes-Querol, 1996). Occult malignancy is a far more common cause of dysphagia than cervical osteophytes and full investigation is necessary in order to exclude this possibility, before attributing the symptoms to the osteophytes. Since this condition is common in the elderly population, radiological demonstration of large osteophytes in a patient with dysphagia should not be presumed to be the cause. These patients should be fully investigated. Radiographical investigations that are useful in the diagnosis are lateral radiograph of the neck, barium swallow and occasionally computed tomography (CT) scanning. Endoscopy may also be indicated. It should be borne in mind that the presence of large osteophytes makes perforation of the oesophagus more likely during rigid oesophagoscopy.

Most of the patients with Forestier's disease are not sufficiently distressed to merit surgery for the vertebral hyperostosis. In dysphagic patients conservative management includes dietary modifications regarding food consistency and swallowing therapy with instructions on neck positioning. Jonathan and Baer (1990) treated one of their patients with Mucaine, with relief of symptoms. Surgical treatment involves removal of the osteophytic overgrowth, either via an anterior cervical approach for C3 to C7 or via the transoral approach for C1 and C2 (Brandenberg and Leibrock, 1986). Following surgical removal osteophytes can recur. Maran and Jacobson (1971), in a review, have suggested anterior cervical fusion after removal of the osteophytic cervical disc in order to prevent osteophyte recurrence, especially when treating younger patients. Other potential complications include vocal fold paralysis (Gamache and Voorhies, 1980), Horner's syndrome, oesophageal perforation and fistula formation (Brandenberg and Leibrock, 1986). In our elderly patient with upper airway obstruction and stridor, a tracheostomy with a speaking valve proved quite sufficient.

Conclusion

Although the condition of airway compromise and/or dysphagia is rare in a patient with osteophytic cervical spine, otolaryngologists may come across similar cases in their practice. These patients should be properly evaluated and treated, preferably with the involvement of different specialists such as radiologists, rheumatologists, gastro-

enterologists, neurosurgeons and orthopaedic surgeons, because of the variety of clinical expression of Forestier's disease.

References

- Barsamian, J. G., Cobb, L. W., Bremer, A. M., Schemer, R. B., Northup, H. M. (1985) Radiographic, clinical and histopathologic evaluation with surgical treatment of Forestier's disease. *Oral Surgery Oral Medicine Oral Pathology* **59**: 136–141.
- Brandenberg, G., Leibrock, L. G. (1986) Dysphagia and dysphonia secondary to anterior cervical osteophytes. *Neurosurgery* **18**: 90–93.
- Deutsch, E. C., Schild, J. A., Mafee, M. F. (1985) Dysphagia and Forestier's disease. *Archives of Otolaryngology* **111**: 400–402.
- Forestier, J., Rotes-Querol, J. (1950) Senile ankylosing hyperostosis of the spine. *Annals of the Rheumatic Diseases* **9**: 321–330.
- Gamache, F. W., Voorhies, R. M. (1980) Hypertrophic cervical osteophytes causing dysphagia. A review. *Journal of Neurosurgery* **53**: 338–344.
- Hassard, A. D. (1984) Cervical ankylosing hyperostosis and airway obstruction. *Laryngoscope* **94**: 966–968.
- Hilding, D. A., Tachdjian, M. O. (1960) Dysphagia and hypertrophic spurring of the cervical spine. *New England Journal of Medicine* **263**: 11–14.
- Jonathan, D., Baer, S. (1990) Cervical osteophytes: their significance in ENT practice (ankylosing vertebral hyperostosis-Forestier's disease). *Journal of Laryngology and Otology* **104**: 236–238.
- Maran, A., Jacobson, I. (1971) Cervical osteophytes presenting with pharyngeal symptoms. *Laryngoscope* **81**: 412–417.
- McCafferty, R. R., Harrison, M. J., Tamas, L. B., Larkins, M. V. (1995) Ossification of the anterior longitudinal ligament and Forestier's disease: an analysis of seven cases. *Journal of Neurosurgery* **83**: 13–17.
- Meeks, L. W., Renshaw, T. S. (1973) Vertebral osteophytosis and dysphagia. *Journal of Bone and Joint Surgery* **55**: 197–201.
- Resnick, D., Niwayama, G. (1976) Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). *Radiology* **119**: 559–568.
- Resnick, D., Shaul, S. R., Robins, J. M. (1975) Diffuse idiopathic skeletal hyperostosis (DISH): Forestier's disease with extraspinal manifestation. *Radiology* **115**: 513–524.
- Rotes-Querol, J. (1996) Clinical manifestations of diffuse idiopathic skeletal hyperostosis (DISH). *British Journal of Rheumatology* **35**: 1193–1196.
- Verstraete, W. L., Cauwer, H. G., Verhulst, D., Jacobs, F. (1998) Vocal cord immobilisation in diffuse idiopathic skeletal hyperostosis. *Acta Otolaryngologica Belgica* **52(1)**: 79–84.

Address for correspondence:

Mr K. Papakostas,
ENT Department,
University Hospital Aintree,
Longmoor Lane,
Liverpool L9 7AL.