The Journal of Laryngology and Otology
June 1987. Vol. **101.** pp. 627–632

Diffuse idiopathic skeletal hyperostosis with dysphagia
(A review)

by

E. Eviatar and M. Harell (Holon, Israel)

Abstract

Dysphagia due to cervical osteophytes is not common. However, diffuse idiopathic skeletal hyperostosis (DISH) with cervical involvement which causes dysphagia is even rarer. The otolaryngologist is not generally familiar with this entity. The diagnosis can be made by plain cervical X-ray films, a barium swallowing esophagogram and or a CT scan of the neck. When doubt still exists, further extra-axial X-ray films can be helpful. Although most patients have been treated surgically, there may be a role for conservative therapy initially, as surgery in elderly DISH patients is often morbid and even fatal.

A 79-year-old patient with DISH (Forestier's disease) is reported. Non-steroidal anti-inflammatory therapy was successfully implemented. DISH is compared with other disorders of the cervical spine which may cause dysphagia.

A Case report

A 79-year-old male outpatient was referred with a progressive dysphagia to solids and moderate pain on swallowing. The patient denied hoarseness, shortness of breath, or trauma to his neck.

On physical examination, the movement of the cervical spine was within the normal range. Indirect laryngoscopy revealed a large mass, approximately 2.5 cm. × 3 cm., protruding from the posterior wall of the hypopharynx. The mucous membrane overlying it was intact. On palpation with a probe, the mass was found to be of bony consistency. Routine blood tests, including glucose and ESR, were normal.

Lateral neck X-ray revealed an elongated bony mass along the first-to-seventh cervical vertebrae. A local osteophyte was found at the height of the sixth cervical vertebra. The widths of the intervertebral spaces were moderately narrowed (Fig. 1). On the barium swallow, mild slowing of the barium column at the height of the sixth vertebra was noted.

A CT scan of the cervical spine revealed an elongated ossification of the frontal planes of the vertebral bodies, especially at the height of the sixth vertebra, with elongated and spiky spurs projecting into the soft tissues of the neck (Figs. 2 and 3).

The patient was treated for five weeks with non-steroidal anti-inflammatory drugs, a stiff collar and soft diet until all symptoms were alleviated. He has been asymptomatic for two years.

Discussion

In 1950 Forestier and Rotes-Querol described for the first time an ankylosing disease of the spine developing in elderly people; this subsequently came to be known as diffuse idiopathic skeletal hyperostosis (DISH). Twenty years later, descriptions of dysphagia resulting from this disease (Resnick and Niwayama, 1976; Resnick *et al.*, 1978) were first published, although Zahn recognised cervical osteophytes causing dysphagia in 1904 (Gamache and Voorhies, 1980). DISH is now known to occur most commonly in middle-aged and elderly males (Forestier and Rotes-Querol; Gamache and Voorhies, 1980; Lambert *et al.*, 1981; Resnick and Niwayama, 1976).

Anatomically, the esophagus is anchored at two points: at the level of the cricoid cartilage and at the diaphragm, while the remainder of the esophagus from the neck to the abdomen is fairly mobile. Thus, at the level of the cricoid a small osteophyte can cause dysphagia. Resnick *et al.* (1975) showed that the fourth through seventh cervical vertebrae

https://doi.org/10.1017/S0022215100102403 Published online by Cambridge University Press



FIG. 1

Lateral cervical X-ray: Contiguous ossification along the first to seventh cervical vertebrae, with a localized osteophyte at the sixth vertebra. The intervertebral spaces are moderate in width.

were the most frequently involved. Abnormalities of the third cervical vertebra were less common, and involvment of the second and first, unusual. Reviews of the pertinent literature by Hilding and Tachdijian in 1960 (36 cases) and by Lambert *et al.* in 1981 (40 cases) indicated that these cervical localisations still held true. (These cases included dysphagia in both cervical spine pathologies in general, and in DISH).

DISH is still unfamiliar to many physicians, although it is common in the general population, having been observed in 6 to 12 per cent of routine autopsy specimens in the axial and extra-axial skeleton (Resnick and Niwayama, 1976; Vernon-Roberts *et al.*, 1974). Involvement of the *cervical* spine is found in 76 per cent of those affected (Resnick *et al.*, 1975). Asymptomatic hypertrophic spurs of the anterior surface of the vertebrae may occur in 20 to 30 per cent of the population (Gamache and Voorhies, 1980).

The number of reported cases of DISH involving dysphagia is small. Lambert *et al.* (1981) reviewed 11 cases, including three cases of their own. In fact, DISH-related dysphagia is probably more widespread, given that many instances were included in orthopedic articles when the dysphagia was one of

https://doi.org/10.1017/S0022215100102403 Published online by Cambridge University Press

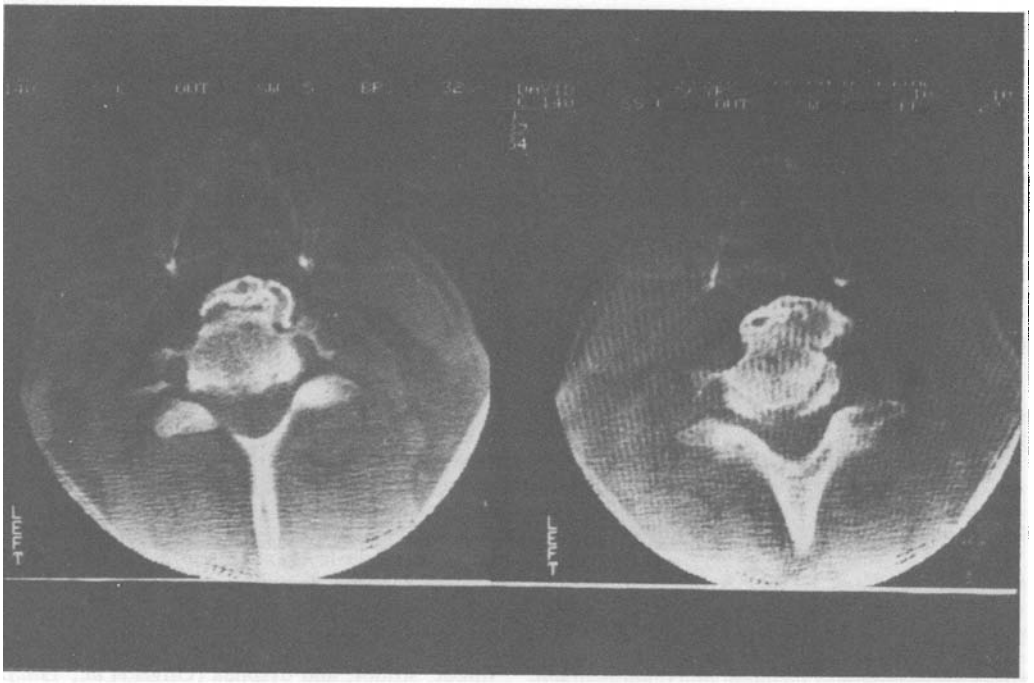


FIG. 2
CT scan of cervical spine: A large osteophyte protrudes into the soft tissues.

the symptoms in the case history (Resnick *et al.*, 1975). DISH is confused with ankylosing spondylitis, spondylosis deformans, and degenerative disc disease.

The differential diagnosis of dysphagia includes such entities as tumours of the neck, mediastinum or spine, esophagitis, esophageal stricture, aberrant vessels, Zenker's diverticulum, neuromuscular disorders, and cricopharyngeal spasm. Lambert *et al.* (1981) suggest that the most difficult differential diagnosis is from gastro-esophageal reflux, when the patient complains of dysphagia originating only in the neck. When anterior cervical osteophytes are present on spinal films, the differential diagnosis suggests congenital bone bars (Gamache and Voorhies, 1980), atlanto-axial spurs, osteochondromas, spondylosis deformans, degenerative disc disease, or anterior herniation of intervertebral disc degeneration.

The etiology of DISH is obscure (Resnick and Niwayama, 1976). It is associated with diabetes mellitus and obesity (Forestier and Rotes-Querol, 1950; Julkunen *et al.*, 1971; Resnick *et al.*, 1975). Toxic exposure to Vitamin A or fluorine has been suggested as an etiologic factor (Resnick *et al.*, 1975).

Pathologically, Forestier and Rotes-Querol (1950) believed that spinal ossification did not originate from the common anterior vertebral ligament; they thought it probable that they arose in the continuous perivertebral fibrous sheaths, though they could not explain why this fibrous vertebral sheath suddenly produced bone. However, Resnick and Niwayama (1976) believe that ossification in the cervical region arises in the ligaments, particularly the ligamentum nuchae. Others speculate that ossification results from decreased mobility, but also, in a vicious circle, is one of its causes. All calcifications and ossifications were early abnormalities associated with chondroid metaplasia and endochondral ossification. In addition, peripheral tears of the annulus fibrosus, anterolateral extensions of fibrous tissue arising from the intervertebral disc, hypervascularity, and chronic inflammatory cells were apparent. Periosteal bony accretion was a conspicuous manifestation associated with irregular bony excrescences on the margins of the vertebral body (Resnick and Niwayama, 1976).

Radiologically, Resnick *et al.* (1975) found preservation of disc height, integrity of apophyseal joints, and absence of outgrowths along the posterior aspects of the vertebral body. Another manifestation of this disease is the tendency for

https://doi.org/10.1017/S0022215100102403 Published online by Cambridge University Press

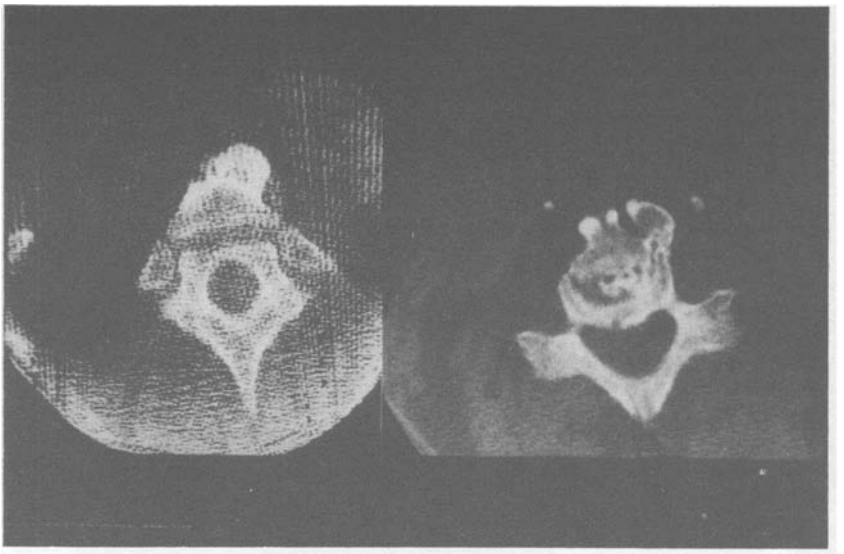


FIG. 3

CT scan of cervical spine: Ossifications along the anterior surface of the vertebra with 'finger-like' appearance.

calcification at sites of ligament and tendon attachments to bone, occurring in extraspinal as well as axial locations. Patellar, olecranon, and calcaneal spurs are also found.

It is unclear how cervical osteophytes cause dysphagia; however, several possible explanations have been advanced: (a) a 'giant' osteophyte may cause dysphagia by simple mechanical obstruction (Girgis *et al.*, 1982; Saunders 1970); (b) a smaller osteophyte may impinge on sites of relative immobility, *e.g.*, the cricoid cartilage area; (c) inflammation in the immediate vicinity of the osteophyte (Lambert *et al.*, 1981); (d) pain and spasm (Umerach *et al.*, 1981). Most probably a combination of some or all of these circumstances is responsible for dysphagia in many cases (Barsamian *et al.* 1985; Lambert *et al.*, 1981).

The most frequent symptom in patients with cervical DISH is dysphagia, although mid or low back pain also commonly occurs is this disease (Resnick *et al.*, 1978).

These authors found dysphagia in 28.6 per cent of the cases histories they reviewed and much less commonly dysphagia was the presenting symptom. Gamache and Voorhies (1980) found it highly probable that the great majority of cases with cervical dysphagia reported in the older literature actually represented patients with DISH.

DISH patients have also complained of odynophagia and/or hoarseness (Barsamian *et al.*, 1985), coughing or the sensation of a lump in the throat, stridor, and dyspnea (Girgis *et al.*, 1982). Dysphagia was gradually progressive in most cases and was always more pronounced with solids than with liquids (Lambert *et al.*, 1981). On examination, a decreased range of motion in the cervical and low back area was the most frequent physical finding (Barsamian *et al.*, 1985; Forestier and Rotes-Querol, 1950; Lambert *et al.*, 1981; Resnick *et al.*, 1975). Indirect laryngoscopy may reveal pharyngeal masses, bony hard on palpation.

By a simple cervical X-ray, the differential diagnosis is reduced to a minimum. Resnick and Niwayama (1976) suggested radiographic criteria for diagnosis: (a) 'flowing' calcification and ossification along the anterior surface of at least four continguous vertebral bodies, most commonly involving the lower thoracic spine; (b) integrity of the disc spaces; (c) normal or mild sclerosis and occasional osteophytes of apophyseal joints, which allows differentiation from ankylosing spondylitis and disc degeneration. Resnick *et al.* (1975, 1978) also described extraspinal radiographic alterations which can help diagnose DISH even without spinal abnormality.

Besides plain X-rays of the spine and extraspinal skeletal regions, most authors recommend barium esophagograms. The findings include a complete stop in the barium column, distortion of the esophagal diameter (Gamache and Voorhies, 1980), a filling defect, or minimal changes when a transient sharp cut-off is seen for a few seconds (Umerach *et al.*, 1981).

https://doi.org/10.1017/S0022215100102403 Published online by Cambridge University Press

The use of computerised tomography and scanning for the diagnosis of DISH is not widespread, but this tool gives us a clear image of the ossification and calcification on the anterior surface of the vertebral bodies, which corresponds well with Resnick's findings (*see* Fig. 3).

When therapy was considered the patient's age and the general consensus in the literature helped us to decide on a conservative therapeutic trial. The treatment of choice for DISH is still in dispute. The range of treatment is wide and even includes radiotherapy (Gamache and Voorhies, 1980), which is not accepted by most authors. Some, *e.g.,* Lambert *et al.* (1981) prefer an initial course of conservative treatment, which includes steroidal and non-steroidal anti-inflammatory drugs with or without antibiotics (Bauer, 1953), to reduce edema without frankly treating the osteophyte itself (Ratnesar, 1970). However, in their review, Gamache and Voorhies (1980) concluded that the procedure of choice is most probably surgical excision of the offending osteophyte, although anterior cervical discectomy is easier (Bone *et al.,* 1974; Iglauer, 1938; Patterson and Byerly, 1958; Saffouri and Ward, 1974). The major risk of this approach is paralysis of the recurrent laryngeal nerve.

The prognosis of DISH is uncertain because only few case reports include adequate follow-up. Three deaths have occurred as a direct result of cervical spine pathology, two died two days post-osteophytectomy (of respiratory arrest) and a third patient died of respiratory failure due to osteophytic compression of the trachea (Lambert *et al.,* 1981; Patterson and Byerly, 1958; Schnier, 1972). Three other DISH patients died of unrelated causes, such as carcinoma of the larynx and cervical esophagus (Gamache and Voorhies, 1980; Heck, 1956: Ratnesar, 1970). Another patient died of myocardial infarction five years after his dysphagia-related DISH was diagnosed (Lambert *et al.,* 1981), and yet another death was caused by fulminating pneumonia three months after diagnosis, without DISH treatment prior to the development of the pneumonia (Girgis *et al.,* 1982).

References

Barsamian, J. G., Cobb, L. W., Bremer, A. M., Scheffer, R. B. and 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.

Bauer, F. (1953) Dysphagia due to cervical spondylosis. *Journal of Laryngology and Otology,* **67:** 615–630.

Bone, R. C., Nahum, A. M. and Harris, A. S. (1974) Evaluation and correction of dysphagia—producing cervical osteophytosis. *Laryngoscope,* **84:** 2045–2050.

Facer, J. C. (1967) Osteophytes of the cervical spine causing dysphagia. *Archives of Otolaryngology,* **86:** 341–345.

Forestier, J. and Rotes-Querol, J. (1950) Senile ankylosing hyperostosis of the spine. *Annals of the Rheumatic Diseases,* **9:** 321–330.

Gamache, F. W. and Voorhies, R. M. (1980) Hypertrophic cervical osteophytes causing dysphagia. A review. *Journal of Neurosurgery,* **53:** 338–343.

Girgis, I. H., Guirguis, N. N. and Mourice, M. (1982) Laryngeal and pharyngeal disorders in vertebral ankylosing hyperostosis. *Journal of Laryngology and Otology,* **96:** 659–664.

Heck, C. V. (1956) Hoarseness and painful deglutition due to massive cervical exostoses. *Surgery, Gynecology and Obstetrics,* **102:** 657–660.

Hilding, D. A. and Tachdijian, M. O. (1960) Dysphagia and hypertrophic spurring of the cervical spine. *New England Journal of Medicine,* **263:** 11–14.

Iglauer, S. (1938) A case of dysphagia due to an osteochondroma of the cervical spine—osteotomy-recovery. *Annals of Otology, Rhinology and Laryngology,* **47:** 799–803.

Julkunen, H., Heinonen, O. P. and Pyorala, K. (1971) Hyperostosis of the spine in an adult population. Its relation to hyperglycemia and obesity. *Annals of the Rheumatic Diseases,* **30:** 605–612.

Lambert, J. R., Tepperman, P. S., Jimenez, J. and Newman, A. (1981) Cervical spine disease and dysphagia. Four new cases and review of the literature. *American Journal of Gastroenterology,* **76:** 35–40.

Patterson, H. A. and Byerly, G. (1958) Esophageal obstruction due to lesions of the cervical spine. *Annals of Surgery,* **147:** 863–867.

Ratnesar, P. (1970) Dysphagia due to cervical exostosis. *Laryngoscope,* **80:** 469–471.

Resnick, D. and Niwayama, G. (1976) Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). *Radiology,* **119:** 559–568.

Resnick, D., Shapiro, R. F., Wiesner, K. B., Niwayama, G., Utsinger, P. D. and Shaul, S. R. (1978) Diffuse idiopathic skeletal hyperostosis (DISH). (Ankylosing hyperostosis of Forestier and Rotes-Querol) *Seminars in Arthritis and Rheumatism,* **7:** 153–187.

Resnick, D., Shaul, S. R. and Robins, J. M. (1975) Diffuse idiopathic skeletal hyperostosis (DISH): Forestier's disease with extraspinal manifestations. *Radiology,* **115:** 513–524.

Saffouri, M. H. and Ward, P. H. (1974) Surgical correction of dysphagia due to cervical osteophytes. *Annals of Otology, Rhinology and Laryngology,* **83:** 65–70.

Saunders, W. H. (1970) Cervical osteophytes and dysphagia. *Annals of Otology, Rhinology and Laryngology,* **79:** 1091–1097.

Schnier, B. R. (1972) Pseudotumor of the hypo-

https://doi.org/10.1017/S0022215100102403 Published online by Cambridge University Press

pharynx and larynx due to anterior cervical osteophytes. *American Journal of Roentgenology,* **115:** 544–546.

Umerach, B. C., Mukherjee, B. K. and Ibekwe, O. (1981) Cervical spondylosis and dysphagia. *Journal of Laryngology and Otology,* **95:** 1179–1183.

Vernon-Roberts, B., Pirie, C. J. and Trenwith, V. (1974) Pathology of the dorsal spine in ankylosing hyperostosis. *Annals of the Rheumatic Diseases,* **33:** 281–288.

Address for correspondence:
M. Harell, M.D.,
Head, Department of Otolaryngology,
The Edith Wolfson Hospital,
Holon 58100, Israel.

https://doi.org/10.1017/S0022215100102403 Published online by Cambridge University Press