

DISH syndrome in ORL practice

St. Stoyanov¹, K. Asenova², P. Kolev¹, Sv. Vasileva¹, Sl. Adamova¹, D. Skerleva³

¹ENT clinic, Ministry of interior - Medical institute, Sofia; ²ENT clinic, Military medical academy, Sofia; ³Kyoto University, Kyoto, Japan



Introduction

Forestier's disease was first described by Forestier and Routes-Querol in 1950. In 1975 was introduced the acronym DISH (diffuse idiopathic skeletal hyperostosis). It is a noninflammatory disease that mainly affects elderly men.[1] Although not rare, it often remains undiagnosed. In the United States, it is found in 25% of men and 15% of women older than 50 years and 35% of men and 26% of women older than 80 years. [2]

The criteria for diagnosis, identified by Resnick and Niwayama are: (1) calcification and ossification along the anterolateral paravertebral ligaments, contiguously involving at least four vertebral bodies with or without specific bony outgrowths projecting into the intervertebral spaces; (2) relative preservation of intervertebral disc height in the involved areas without signs of degeneration; and (3) absence of apophyseal ankylosis or erosion/sclerosis/sacroiliac fusion. [3] Typically, osteophytes are identified in the anterior and lateral regions of the vertebral bodies. Thoracic spine is most frequently affected, followed by the lumbar and cervical regions. [1-3]

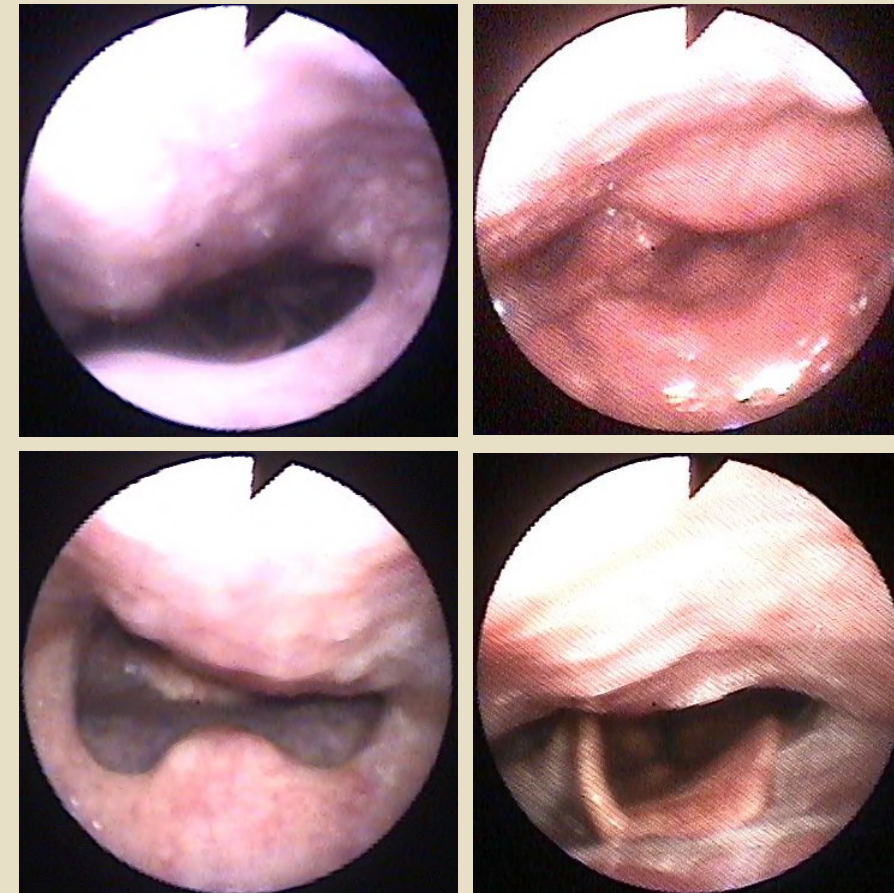
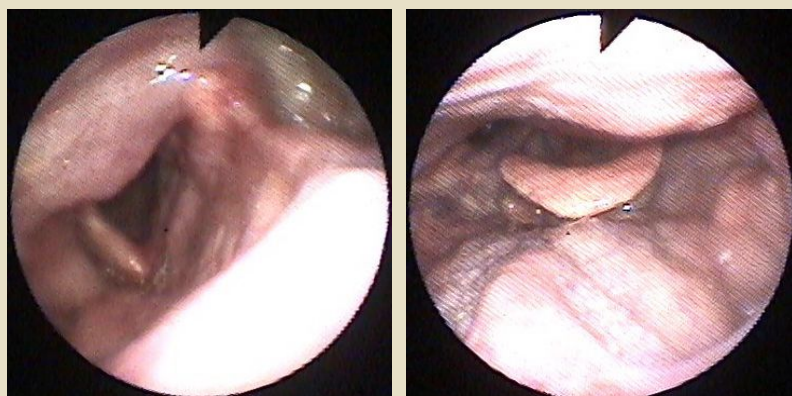
DISH is usually asymptomatic. [1] In the cervical region, it can generate ENT symptoms as dysphagia, pharyngeal globus, dysphonia, and stridor. [5] Dysphagia is the most frequently reported symptom and can occur in up to 28% of cases, and be directly related to the presence of cervical osteophytes. [5] It is usually progressive and more severe for solid foods than liquids. Other symptoms are cough, sore throat, foreign body sensation and sleep apnea. [3,4]

Objective

We would like to share our experience with DISH syndrome patients with anterior cervical ligament involvement. They may have swallowing problems and sometimes even airway obstruction. The posterior pharyngeal wall swelling on laryngoscopy may raise concern for malignancy.

Methods

We studied 14 patients (9 – male and 5 – female), aged between 62 and 94 years, all of which had visual signs of DISH syndrome on fiberoptic laryngoscopy (fig. 1-6) at the ENT clinic of the Medical institute of Ministry of Interior



Results

The etiology of DISH remains unclear. [1,3] Initially, trauma was suggested as the precipitating factor, although not present in all cases. [3] The following were indicated as risk factors: excessive mechanical stress with or without obesity, dyslipidemia, hyperuricemia, hypertension, cardiovascular disease, hypervitaminosis. [4,6]

Symptoms of complications are more severe than those of the disease itself. [3] Dysphagia is the best-known cervical symptom, occurring in approximately one-third of patients. [6,7] We found it in 5 of our patients (4-male, 1-female). One of them was with worsening of symptoms in cervical extension and improvement with flexion. [8]

There was no clear correlation between the size of osteophytes and severity of dysphagia, which may be due to the presence of underlying presbyphagia. [3,4,9]

The mechanisms for dysphagia in DISH suggested in the literature include: 1) incomplete protection of the lower airways due to restricted epiglottis mobility; 2) incomplete glottal closure due to restricted vocal fold mobility; 3) restriction of the movement of elevation and anterior displacement of larynx; 4) neuropathy due to impairment of the recurrent laryngeal nerve; 5) mechanical obstruction of food bolus transportation in the posterior wall of the hypopharynx or esophagus; 6) inflammation and fibrosis of the esophageal wall secondary to irritation by osteophytes; and 7) periesophageal spasm due to pain. [2,3,4,7-9]

Patients may rarely have dyspnea (1 of our patients), or even the need for an emergency airway. [3,5,10] Dyspnea can be explained not only by the mechanical obstruction in the airways, but also by retrocricoid inflammation generated by osteophytes, which leads to reduction in glottal mobility. This mechanism also explains the presence of dysphonia and stridor. Osteophytes at the level of C2 and C3 vertebrae can generate higher risk for airway impairment in Forestier's disease. [10] There is also a report of difficult intubation in a patient with DISH [11] and cases of aspiration pneumonia. [12,13]

Reflex otalgia can occur by reflex stimulation of the pharyngeal plexus through the glossopharyngeal and vagus nerves.[2] Medullary compression symptoms occur when there is extension of calcification into the medullary canal or ossification of the posterior longitudinal ligament. [2,14]

Maseiro et al. have suggested a classification for the degree of posterior pharyngeal wall compression. [15] Compression is considered slight when the pharyngeal lumen reduction is less than 30%; moderate, when the reduction is between 30% and 50%; and severe when the reduction exceeds 50%. Most of our patients were with mild to moderate compression.

Radiological imaging is important. Plain radiography of the spinal column is usually sufficient (fig.7), but CT and MRI may be helpful for assessment of affected soft tissue and intramedullary involvement [9]

Differential diagnosis of DISH includes ankylosing spondylitis and spondylosis deformans, degenerative diseases of the intervertebral discs, and osteoarthritis. [1] Acromegaly, fluorosis, pachydermoperiostosis, and hypertrophic osteoarthropathy are less common causes that should also be ruled out. [2] In cases with dysphagia, the possibility of a malignancy should not be overlooked, even if there is radiological evidence of osteophytes. [4] Tumors of the larynx, esophagus, lung, mediastinum, and spinal column should be ruled out. Disorders of esophageal mobility, esophagitis, esophageal stricture, vascular abnormalities, Zenker's diverticulum, Plummer-Vinson syndrome, gastroesophageal reflux, globus hystericus, CVA, neurological disorders, dermatomyositis, isolated ventral cervical osteophytes, elongation of the styloid process of the skull base or calcification of the stylohyoid ligament are other causes of dysphagia that should be kept in mind. [2,4]



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