Respiratory Distress and Dysphagia Secondary to Diffuse Idiopathic Skeletal Hyperostosis: Case Report and Review

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ABSTRACT

Diffuse idiopathic skeletal hyperostosis (DISH) is a common disorder among the elderly. The bridged ossified anterior longitudinal ligament might infrequently cause swallowing difficulties and respiratory distress secondary to airway obstruction. The latter complication is very rare. It is usually due to excessively enlarged cervical osteophytes at the C3 to C4 level with compression of posterior pharyngeal wall and upper airway leading to edema, partial obstruction or distortion of the laryngeal inlet and consequent dyspnea. Herein a 59-year-old man, with the chief complaints of dysphagia for two years and laryngeal stridor, snoring and hoarseness for a year is presented. He also complained of progressive troublesome respiratory distress during the last three months. Radiological findings were compatible with DISH From C2 To C5 with the largest one at C3-C4. Surgical excision of the prominent osteophytes resulted in improvement of both the dysphagia and respiratory discomfort.

KEY WORDS: DISH, Dyspnea, Dysphagia, Forestier's disease, HACO, Respiratory discomfort

INTRODUCTION

Diffuse idiopathic skeletal hyperostosis (DISH) or Forestier's disease was first described by Forestier and Rotes Querol in 1950. Later in 1975, Resnick et al described diffuse idiopathic skeletal hyperostosis (DISH) (21). Nowadays, DISH is used for the more generalized form of the disease with axial and extraspinal ligaments affection, while Forestier's syndrome is used for cervical manifestation of the condition (2). Based on radiological surveys of the cervical spine 2.4-5.4% of individuals older than 40 years of age have Forestier's disease, although most of them remain asymptomatic throughout the life (28). Neurological manifestation is absent in the patients affecting by this syndrome unless it coexists with OPLL (1). Actually, the most common clinical picture of this disease is dysphagia, and about 3% to 20% of adults are expected to develop swallowing disorder due to this syndrome at some time point in their lives (2,3,6,9,17).

A quite rare clinical picture of the disease is respiratory disturbance resulting from compression or distortion of the airway apparatus by large osteophytes.

A wide spectrum of respiratory disturbances ranging from stridor, hoarseness, sleep apnea and dyspnea to acute respiratory distress have been reported with DISH (3,4,6,8-16,20,22,24-27).

A review of the literature revealed that in the majority of reported cases airway compromise was due to upper cervical osteophytes located through C2 to C4 that could compress both the posterior pharyngeal wall and upper airway corridor (4,6,9,16).

Herein a 59-year-old male with a chief complaint of dysphagia for two years and respiratory distress of increasing intensity for a year is presented. The dyspnea became more evident in recent months occurring both in an effort and during sleep. Radiographs, CT scan
and barium swallow confirmed that it has resulted from pharyngo-laryngeal distortion due to its encroachment by several large contiguous osteophytes more voluminous at the C3-C4 level compatible with Forestier's syndrome. With excision of the corresponding osteophytes, both complaints dramatically improved.

**CASE REPORT**

This 59-year-old male was admitted because of dysphagia of two years duration. He had stridor, hoarseness and sleep apnea for a year that was worsened and changed to dyspnea in the last three months. Dyspnea was in particular troublesome during sleep and with extension of the neck. Respiratory problems had resulted in sleep disturbance that obliged him to sit in the bed for a while several times during sleep hours.

Plain Lateral cervical radiographs disclosed massive ossification of the anterior longitudinal ligament compatible with Forestier's disease. The ossification involved this ligament through the second to fifth cervical vertebra, with the largest one at C3-C4 level causing obstruction of the airway corridor at laryngeal inlet (Figure 1).

Axial CT revealed prominent osteophytes more prominent on the right side. These osteophytes resulted in significant displacement of soft tissues (Figure 2).

Reconstructed CT disclosed osteophyte from C2 to C5 with the most voluminous one at C3-C4 level. There were layers of incomplete ossification anterior to these osteophytes at this segments. This clearly demonstrates the actual size of the osteophyte that was two times larger than what was seen in plain radiographs (Figure 3A,B).

Barium swallow showed marked distortion of the pharyngo-esophageal segment to the right by an extremely large bony mass (Figure 4).

Endoscopy was regarded contraindicated in this particular patient because of the associated severe distortion.

Since intubation was postulated to be difficult and dangerous, even with fiber optic assistance, preoperative tracheotomy was done with application of local anesthetics...
and sedation. Later, general anesthesia induction was accomplished through the tracheotomy tube. Then, a right-sided incision along the anterior to the border of sternocleidomastoid muscle was made. Later with blunt dissection of the soft tissues, large osteophytes from C2 to C5 were exposed which were removed with high-speed

![Figure 3: A) Axial reconstructed C7-T1 Images showing obstruction of upper airway. Note the layer of incomplete ossification anterior to the osteophytes. B) Schematic drawing of reconstructed CT disclosing the events and the pattern of obstruction.](image)

![Figure 4: Barium swallow demonstrating severe distortion of upper pharynx around a dense bony mass compatible with asymmetrical osteophytes.](image)

![Figure 5: Post-operative lateral cervical X-ray taken a year after resection of the osteophytes.](image)
cutting burr. Subsequently a diamond match stick was used to smooth the region.

The patient was transferred to ICU under controlled ventilation. Next day, with discontinuation of anesthetic medications, the patient became fully alert. Then, the machine was switched to assisted ventilation and later was he weaned to spontaneous mode. Two days after surgery, the tracheotomy tube was capped in order to allow him to attempt normal breathing and it was removed subsequently.

On the fifth operative day, he was discharged with no dyspnea or swallowing difficulty anymore. A year after surgery, he had no complaint and lateral cervical X-Ray did not show any osteophyte (Figure 4).

**DISCUSSION**

DISH is a non-inflammatory disorder of unknown etiology which is common among the elderly having potential associations with obesity, related metabolic conditions, and high social status (9,21). There is a statistical correlation between vertebral hyperostosis and Dupuytren’s contracture in men beyond the age of 60 (9). The condition can be recognized in plain lateral cervical radiograph with the presence of “flowing candle-wax” ossification along the anterolateral margins of at least four contiguous vertebral bodies. Disc height remains normal in this condition and with minimal affection of the cervical apophyseal joints (6,9,21).

Similar changes are demonstrated in hypertrophic anterior cervical osteophytes (HACO) where discs height are decreased due to cervical spondylosis. Clinically hypertrophic cervical osteophyte might mimic DISH (9). DISH is more common in men and occurs primarily in the elderly population and might be associated with the syndromes of joints osteoarthritis and ossification of the posterior longitudinal ligament (15,21). Neurological deficits are rare in DISH and only occur where ossification of posterior longitudinal ligament coexists (1). However, dyspnea and dysphagia might be seen in different intensities.

Anatomically, below C4, esophagus lies in front of the vertebral column and anterior to the esophagus is the trachea. Osteophytes below C4 can cause moderate to severe dysphagia as the result of direct esophageal compression. Therefore, DISH with prominent osteophytes below C4 should be regarded as uncommon but well-known cause of dysphagia in particular in the elderly (2,9,17). However, ossifications at these locations rarely cause of dyspnea. Esophageal compromise might be followed by compression of the trachea that eventually causes respiratory discomfort where an excessively large osteophytes is necessary for this complication (26). However, besides direct compression of the trachea, direct entrapment and affection of both laryngeal nerves with bilateral vocal cord immobility or paralysis and emphysema secondary to perforation of trachea are other postulated rare causes of respiratory distress (11). With osteophyte locating at the borderline region of C4-C5, dyspnea may not be only due to narrowing or obstruction of the airway, but might be due to retrocricoid inflammation and necrosis that can lead to reduced vocal cord mobility (11,15). This disorder may result in dysphonia, stridor and in severe cases aspiration with acute respiratory distress where urgent tracheotomy is required. Further, in one rare occasion, combination of the compressive effect of enlarged sternoclavicular joint and osteophyte at C7-T1 resulted in severe compression of tracheoesophageal systems circumferentially.

Above C4, the vertebral bodies are in touch with pharynx where large osteophytes through C2 to C4 may result in dysphagia and respiratory distress (4,6,10,16). Initially, the patient complains of the sensation of a retropharyngeal foreign body or globus and later true dysphagia.

At this region, respiratory distress secondary to DISH is regarded to be due to partial obstruction of the upper airway corridor secondary to combination of direct extrinsic compression from the osteophytes and internal compromise from a reactive intrinsic inflammation (8,9,13,26). If the corresponding osteophyte is larger on one side, it might result in distortion of upper aerodigestive corridors where it aggravates the preexisting symptoms.

With regard to anatomy, respiratory discomfort at C2-C3 osteophyte is due to impingement of epiglottis and osteophyte at C3-C4 result in compromise of subglottic space and laryngeal inlet (3,4,6).

Effort dyspnea, dyspnea during sleep, stridor, continuous snoring, sore throat and sleep apnea are the various patterns of respiratory distress (3,4,6,8-16,20,22,24-27).

Actually, in the presence of distortion, the pattern of respiratory discomfort develops in faster and more
prominent fashion. Notably, the upper airway obstructive symptoms are usually aggravated with neck extension.

Nonetheless, while dysphagia due to compressive effect of large osteophytes in are published in increasing frequency, reports on respiratory compromise secondary to this pathology are rare and less than 20 cases have been found in the literature since the first report by Benhabyles in 1970 (3,4,6,8-16,20,22,24-27).

Further, to the best of our knowledge, distortion of the upper airway apparatus as a causative mechanism in respiratory disturbance has not been published previously. Therefore, the current case besides its rarity seems the first example that dyspnea has been due to such a severe distortion.

Plain radiographs are necessary for diagnosis of DISH. The diagnostic criteria are ossification within the anterior longitudinal ligaments of at least four contiguous vertebral bodies; preservation of disc space height, and normal or only mildly sclerotic apophyseal joint (2-4,6-17,19,20,22,24-27).

A similar condition which might result in dysphagia and dyspnea is hypertrophic anterior cervical osteophytes (HACO) where the disc height are usually narrowed and apophyseal joints of the cervical spine might be affected.

Mostly, there is a direct relationship between swallowing pathology and, respiratory distress with the size of the osteophytes. Usually with an osteophyte larger than 10 mm, the possibility of the abovementioned complaints is more.

Nonetheless, with the use of X-Ray, impingement and reduced caliber of the aerodigestive tract can be demonstrated. Usually, the constricted tract is located far anterior from the osteophyte (15). Actually, anterior to the osteophytes, there might be several layers of incomplete ossification hardly visible in plain radiographs. Further, intrinsic inflammation anterior to these layers will result in significant aerodigestive tract compromise (15).

With axial and reconstructed sagittal CT images, one can precisely measure the size of the osteophytes and demonstrate the side of projection (3,9,15). The true distance between the osteophytes and the pharyngolaryngeal tract as well as the layers of incomplete ossification can be demonstrated.

Evaluation of swallowing with videofluoroscopy allows a definite diagnosis of dysphagia to be made. Barium swallow can show the site of impingement and should be included in evaluation of such patients (3,9,15,25).

Flexible endoscopy can be used but it must be performed carefully because of the risk of trauma due to the osteophytes (8,9,15,16).

Generally, treatment of DISH or HACO is only indicated when it becomes symptomatic.

With respect to conservative treatment, changing the diet measures and NSAIDs are highly useful for managing the swallowing problems of the elderly patients with often major comorbidities (3,15,25). Most often, medical management is initiated with hygiene and diet modifications, ideally with the assistance of a dietician capable of defining the patient's calorie needs. Mixed, soft, semiliquid foods or the use of food complements are highly useful solutions (2,6). However, when the general health is too precarious, feeding with percutaneous endoscopic gastrostomy (PEG) should be considered (9,25).

It should be noted that conservative therapeutic strategy is less effective in respiratory disturbances (15). In those elderly subjects with serious comorbidities harboring acute or chronic respiratory distress, permanent tracheostomy is beneficial (3,15,25).

Optimal therapeutic strategy for younger patients with dysphagia and dyspnea is surgical removal of the osteophytes (3,9,15). Anterolateral surgical approach with an incision along the anterior border of sternocleidomastoid muscle is recommended for the osteophytes below C3 (9). The side of approach is up to the surgeon. The exception is where the osteophytes are more prominent on one side which eventually dictate the side of incision. Nonetheless, through a long incision and with adequate exposure, the osteophytes can be excised preferably with high-speed cutting burr (9). Subsequently with high-speed diamond match stick, one can smooth the site of pathology. This is true for the elderly with serious airway compromise but without comorbidities where surgical excision of the osteophytic masses is the treatment of choice.

However, a retropharyngeal corridor is recommended for the osteophytes above C3, at the C1-C2 segments (23).

Anesthesia is a very important issue in patients with dysphagia or dyspnea suffering from DISH. In cases with moderate size osteophytes, fiberoptic-assisted intubation might be beneficial (5,19). In those with large osteophytes
or marked distortion, preoperative tracheostomy before induction of general anesthesia is advised (5,15).

In cases with bilateral vocal cord paralysis resulting in aspiration and acute respiratory distress, primary tracheostomy is necessary to reopen the airway (3,15,19). Surgery should be postponed till the general condition becomes stable.

Post–operatively, it is preferred that the patient spends at least one night in ICU under assisted ventilation. A day or two after surgery one can wean the assisted ventilation, cap the tracheostomy tube and remove it (16,25).

Two points are advised: one is elevation of the head and neck to ease venous flow to the heart and the next is pouring Dexamethasone locally at the corresponding territory intraoperatively or its parenteral application post-operatively in the dose of 4 mg three times daily for 48 hours both of which are supposed to decrease postoperative edema.

Surgery can eliminate the symptoms caused by the compressive effects of the osteophytes, except those who sustain irreversible conditions. Therefore, the outcome for dysphagia is mostly good except in those with longstanding history where fibrosis has replaced inflammation (7,8,15,17,19). Prognosis of dyspnea after osteophytectomy has been regarded acceptable to excellent in the majority of the reported cases unless it is due to bilateral vocal cord immobility or paralysis (9,15,16,18). Less satisfactory results might be expected in sleep apnea of the elderly (8). After successful surgical resection, radiological reappearance of DISH with new osteophyte formation in an average rate of 1 mm/year has been reported (18). However, most patients with such a recurrence remain asymptomatic (18).

In conclusion, respiratory distress secondary to DISH is a treatable condition. Airway compromise is usually due to large osteophytes through C2 to C4. With appearance of respiratory compromise, surgery should be undertaken as soon as possible. Preoperative tracheotomy might be necessary before resection of the osteophytes.

REFERENCES


