

[CASE REPORT]

Management of Airway Obstruction due to Diffuse Idiopathic Skeletal Hyperostosis in the Cervical Spine: A Case Report and Literature Review

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Abstract:

Diffuse idiopathic skeletal hyperostosis (DISH) is a relatively common progressive noninflammatory entheses disease. Patients are often asymptomatic or are undiagnosed due to minor chronic symptoms. We herein report a rare case in which the primary symptom was sudden-onset upper airway obstruction due to exuberant osteophytosis in the cervical spine. Treatment was successful with careful airway management and surgical osteophyectomy. Most DISH cases in the literature with airway obstruction have been managed with tracheotomy. However, the safety and necessity of this approach remain questionable. We herein discuss the possibility of conservative management as a choice of airway control. Airway obstruction due to DISH may be underrecognized. This highlights the importance of including DISH in the differential diagnosis of airway obstruction. In addition, a detailed evaluation and personalized care for each individual case is essential.

Key words: diffuse idiopathic skeletal hyperostosis, airway obstruction, respiratory failure, dysphagia, surgery

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Introduction

Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier's disease, was first described by Forestier in 1950 as senile ankylosing vertebral hyperostosis (1). It is a progressive noninflammatory entheses disease, relatively common in elderly men (2), and characterized by the ossification of ligaments and entheses in the prevertebral regions of the spine (3). Its prevalence is said to range from 2.9% in Asians to 25% in Caucasians (4, 5). It is expected to increase in frequency in the future, as the etiology is attributed to metabolic factors, including diabetes mellitus, obesity, and old age. Patients are often asymptomatic or are undiagnosed due to minor chronic and unrevealing symptoms, such as back pain or spinal stiffness (6). Some patients may present with more severe symptoms, such as dysphagia,

spine fractures, and in rare cases, dyspnea (7).

We herein report a rare case of DISH in which the primary symptom was sudden-onset upper airway obstruction, complicated by negative-pressure pulmonary edema. Treatment was successful with careful airway management and surgical osteophyectomy followed by prolonged rehabilitation for dysphagia. We also present a literature review of cases of respiratory failure due to DISH.

Case Report

An 80-year-old man was transported to our emergency department due to respiratory failure. He had developed a productive cough one week before and a sore throat the day before. On the day of onset, exactly when he bent his neck forward to get up from bed, he suddenly could not breathe or talk. He stomped the floor to call for help and was found

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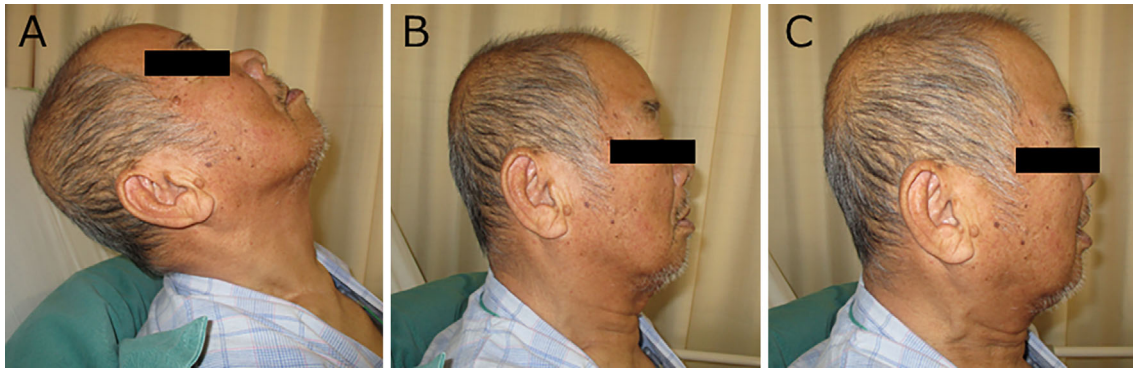


Figure 1. The neck range of motion was quite limited. **A:** maximum extension, **B:** neutral, **C:** maximum flexion.

by his family to be in severe distress. He had a history of cerebral infarction 15 years earlier that left him with slight right leg paralysis and a left testicular tumor treated with no relapse. He also had a 15-year history of type 2 diabetes, hypertension, and hyperlipidemia and was taking aspirin, glimepiride, linagliptin, amlodipine, and atorvastatin. He had never smoked and did not drink alcohol regularly. He had no family history of cardiac or respiratory disease.

On examination, he was alert and in severe distress. He was unable to vocalize and complained of difficulty breathing when bending his neck forward or backward. His oxygen saturation was 90% on 10 L O₂/minute, and his respiratory rate was 24/minute, blood pressure 165/92 mmHg, heart rate 110/minute, and body temperature 36.1°C. His neck range of motion was quite limited, especially when bending forward (Fig. 1). His palatine tonsils were swollen. There was no jugular venous distention or cervical lymphadenopathy. Stridor and wheezes were heard in the neck and all lung fields on inspiration. The heart sounds were normal. There was no skin rash. No new neurological findings were present. Laboratory data were unremarkable (white cell count 9,910/mm³, C-reactive protein 0.74 mg/dL, brain natriuretic peptide 75.5 pg/mL, blood glucose level 157 mg/dL, HbA1c 6.1%).

Chest X-ray and computed tomography (CT) showed diffuse ground-glass opacities bilaterally that were more pronounced in the upper lung fields than the lower lung fields (Fig. 2A-C). There was no heart enlargement or pleural effusion. In the emergency room, his breathing and phonation improved promptly with suctioning and positioning, and supplemental oxygen was lowered to 2 L O₂/minute. Findings were inconsistent with anaphylaxis, congestive heart disease, cerebrovascular disease, or other common causes of respiratory distress. He was admitted for further evaluation and respiratory control.

Ceftriaxone was administered for possible airway infection. The next day, his dyspnea and stridor had improved, and the lung ground glass opacities had disappeared on X-ray (as confirmed by CT performed to evaluate upper airway obstruction), suggesting that the findings had been due to negative-pressure pulmonary edema (Fig. 2D-F). Cervical X-

ray and CT revealed abnormal ossification of the anterior longitudinal ligament from the second to seventh cervical vertebrae (Fig. 3A-C). The bony changes of the anterior ligament of the cervical vertebrae showed a “flowing candle wax” image, typical in cases of DISH. The patient’s demographics (old age, complicated with diabetes), the absence of sacroiliac joint involvement, and these characteristic image findings were inconsistent with ankylosing spondylitis. The thoracic vertebrae involvement was inconsistent with spondylosis deformans. Three-dimensional CT and bronchoscopy revealed severe pharyngeal and tracheal stenosis (Fig. 3D and E, 4). Based on these findings, the patient was diagnosed with DISH.

Increased airway secretion and airway mucosal edema due to airway infection had presumably caused his chronically narrow airway to be narrowed further and finally be completely obstructed when bending his neck forward to get up in the morning, causing negative-pressure pulmonary edema. Surgical decompression was considered necessary due to the high risk for recurrence of airway obstruction. His dysphagia was also problematic and found to be due to restricted laryngeal suspension and esophageal stenosis on a videofluoroscopic swallowing study (VFSS). This was also considered an indication for surgical treatment.

After cessation of aspirin, surgical osteophylectomy (surgical removal of an abnormal bone formation) was performed on day 11 of admission. A percutaneous cardiopulmonary support device was prepared in case of difficult intubation or airway obstruction upon cervical retroflexion for intubation. However, bronchoscopy-assisted intubation with a 5.5-mm spiral tube was successful without cervical flexion. The surgery was uneventful. Ossification of the anterior longitudinal ligament from C3/4 to C6/7 was planarized (Fig. 5A).

Due to the risk of postsurgical hemorrhaging and secondary airway stenosis, the patient was not extubated immediately after surgery. Careful observation and evaluation revealed no clinically significant hematoma, and the patient was extubated the next morning. Improvement of tracheal stenosis was confirmed on three-dimensional CT of the respiratory tract (Fig. 5B and C), and no respiratory distress was noted at any cervical position. His respiratory condition

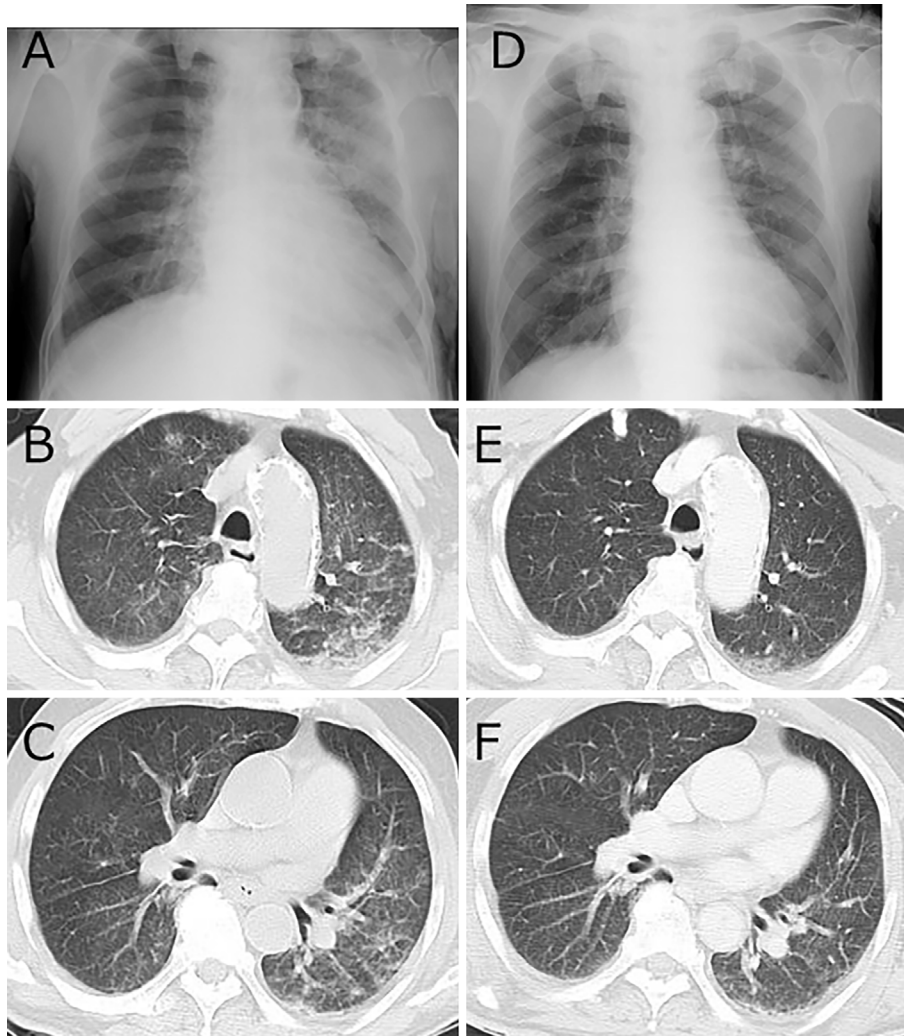


Figure 2. Chest X-ray (A) and CT (B, C) on admission showed bilateral ground glass opacities. On day 2, the bilateral ground glass opacities had disappeared on chest X-ray (D) and CT (E, F). Both X-rays were taken in the anterior-posterior position.

and dysphonia recovered soon after surgery and remained stable throughout the clinical course. However, his dysphagia did not improve immediately. This was attributed to the thickened pharyngeal soft tissue due to chronic stimulation by the protrusion of the ossified ligament, functional decline due to chronic structural abnormality, other possible causes of dysphagia (i.e. cerebral infarction and aging), and repetitive episodes of cholecystitis and colitis delaying aggressive rehabilitation or adequate enteral nutrition. Multi-disciplinary discussion was repeated, and a long-term goal was set to improve the dysphagia.

To improve his general condition, nutrition was provided parenterally from a nasogastric tube, and on day 43 of admission, gastrostomy was performed. Oral care and indirect swallowing training (training without the use of foods or liquids) were performed regularly. A VFSS was repeated six times to assess the swallowing function in a timely manner and provide adequate rehabilitation. Training with small amounts of thickened liquids and food was finally initiated from day 53. He was discharged at four months while eating three meals per day, and the gastrostomy was closed five

months from the onset, at which point he could safely swallow unthickened liquids. Twenty months later, he has not had any recurrent episodes of respiratory distress or aspiration pneumonia.

Discussion

We experienced a rare case of DISH presenting with sudden-onset upper airway obstruction that was managed without tracheotomy. DISH is a progressive noninflammatory disease characterized by ossification (bone tissue formation) of the anterior part of the vertebrae (8). It develops chronically and commonly presents with nonacute symptoms, such as back or neck pain, spine mobility restriction, foreign body sensation, and hoarseness. Dyspnea may occur in some cases but rarely has been reported to occur suddenly. In a review article of literature from 1980 to 2010, a total of 189 cases with dysphagia and 63 cases with airway obstruction (including 48 with both) were reported (9). In our case, the presenting symptom was sudden-onset airway obstruction. The patient had experienced symptoms suggest-

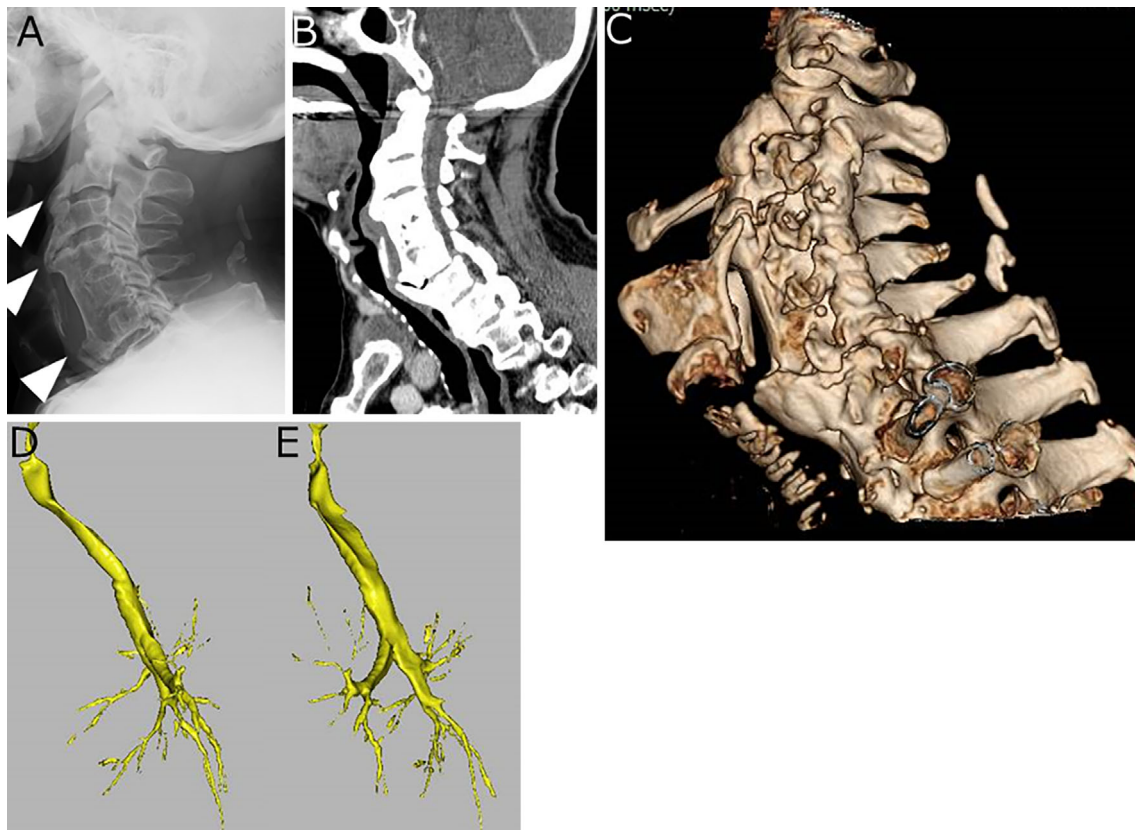


Figure 3. Cervical X-ray (A), CT (B), and three-dimensional CT (C) showed abnormal ossification (arrowheads) of the anterior longitudinal ligament from the second to seventh cervical bones. Three-dimensional CT of the respiratory tract (D, E) revealed severe tracheal stenosis.

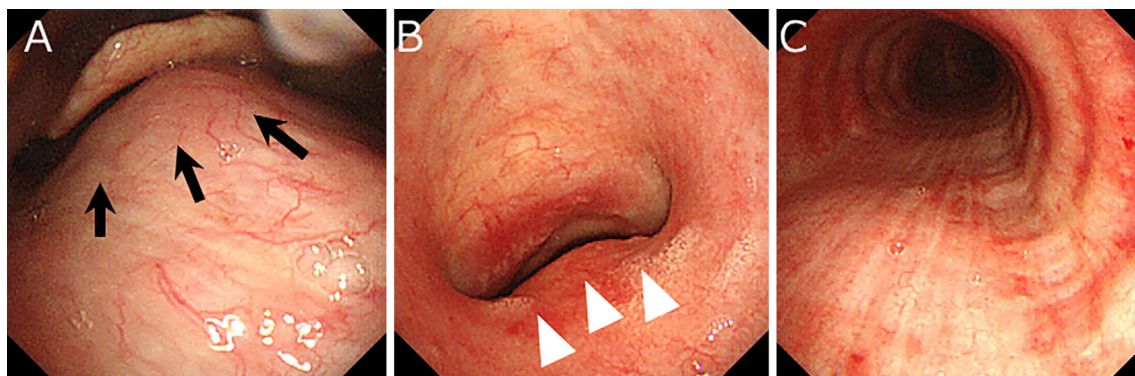


Figure 4. Bronchoscopy images. A: Pharyngeal stenosis due to cervical protrusion (arrows). B: Tracheal stenosis due to cervical protrusion (arrowheads). C: The trachea peripheral of the cervical protrusion was intact.

ing an upper respiratory infection from a few days before onset. This may have caused edematous changes to the upper respiratory membrane. The airway obstruction occurred exactly when he bent his neck forwards, indicating that the neck bending was the final trigger in obstructing the narrow and edematous airway. Airway obstruction was reproducible with neck bending, which also supported this diagnosis. Similar reports included a case of airway obstruction due to DISH causing negative-pressure pulmonary hemoptysis (10). The reported case required intubation due to respiratory failure and loss of consciousness.

The airway management of DISH affecting the airway is important. In the above-mentioned report, 20 of the 24 cases with severe airway obstruction underwent emergency intubation or tracheotomy (9). In our case, emergency intubation or tracheotomy was also considered. However, attempts at intubation may have worsened the airway mucosal edema. Tracheotomy may have caused uncontrollable hemorrhaging since the patient had been taking aspirin. It also may have caused additional complications due to surgery and cannulation. We therefore decided to manage him conservatively while discontinuing aspirin and preparing for surgical osteo-

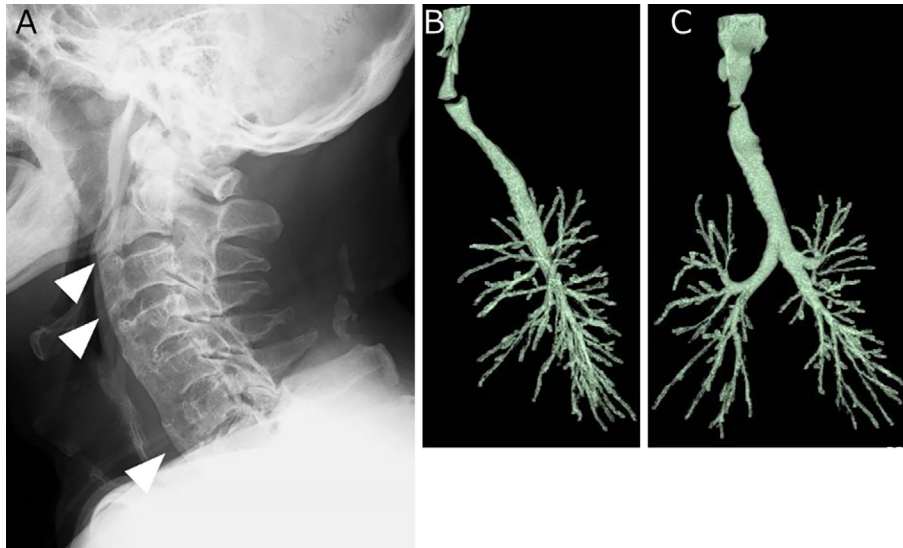


Figure 5. Post-surgical cervical X-ray (A) showed that ossification of the anterior longitudinal ligament from C3/4 to C6/7 was planarized (arrowheads). Post-surgical three-dimensional CT of the respiratory tract (B, C) showed that the tracheal stenosis had improved.

phyectomy. Careful positioning of the neck at the patient's most comfortable angle and gentle suctioning of the airway secretions at the appropriate timing were effective, and his airway obstruction improved overnight. Perioperative airway management was also safely performed without tracheotomy. Nelson et al. pointed out that airway obstruction can be the initial presentation with laryngeal findings that might not be explained purely by mechanical obstruction (11). Instead, the airway obstruction may be caused by a secondary inflammatory response in the postcricoid area or fixation of vocal cords. Inflammation and possible reflux as well as aspiration in addition to the already narrow airway (due to osteophytes) can readily cause airway obstruction, as in our case. Therefore, improvement of reversible factors (inflammation, reflux, and aspiration) may be the key to safe airway management of these patients. In our case, we focused on treating the possible infection with antibiotics, and prohibited any oral intake in order to prevent further aspiration. The upper body was kept raised to prevent reflux. The utmost care was taken to maintain the neck position, especially during X-ray examinations and bronchoscopy. In addition, some reports have described the usage of corticosteroids, non-steroidal anti-inflammatory drugs, antireflux drugs, and gastroprokinetics (9). The intake of soft foods and fluids and the application of a soft cervical collar have also been reported. These conservative and supportive measures may all be important for managing DISH with airway obstruction.

While physicians should always be prepared to perform tracheotomy, they should also bear in mind that aspects of the procedure itself, such as neck distention and careful palpation, can directly cause airway obstruction. Furthermore, routine tracheotomy procedures may be difficult due to osteophytes. It may be necessary to select a longer tube in order to perform safe intubation over the narrowed portion of

the airway or to adopt a lower position for the incision. In our case, we had cardiovascular surgeons on standby prepared to perform extracorporeal circulation if necessary. The most important factor in the treatment of such cases seems to be for the multidisciplinary staff to anticipate possible problems and engage in active discussion in order to be prepared to adopt any of several management strategies.

DISH can occur in any part of the human skeleton; however, symptoms due to cervical ossification can be the most life-threatening. Through CT analyses of DISH and healthy subjects, Bakker et al. found that hyperostosis at the cervical level was symmetrically distributed anterior to the vertebral bodies without a flowing pattern, in contrast to the asymmetrical flowing pattern typically found in the thoracic spine (12). The significant ventral displacement of the trachea and esophagus may be involved in the mechanism underlying the dysphagia and airway obstruction observed in DISH.

In a retrospective review of 234 patients encountered at a single institution over a 20-year period with the diagnosis of cervical osteophytes, 9 (3.8%) presented with dysphagia (11). Three of these nine patients developed acute airway obstruction and required intubation and tracheotomy.

Airway obstruction due to DISH has been considered rare; however, considering the recent incidence of DISH itself and the increasing frequency of predisposing factors (old age, diabetes mellitus, etc.), it may be underrecognized. The present case highlights the importance of including DISH in the differential diagnosis of airway obstruction. In addition, to enable safe airway management in cases of DISH, a detailed evaluation and personalized care for each individual patient is essential.

Conclusion

We experienced a unique case of DISH presenting with

sudden-onset airway obstruction as the primary presentation that was managed safely with conservative measures initially. DISH is a common but underdiagnosed condition. This case highlights the importance of including DISH in the differentiation of acute respiratory distress.

Informed consent was obtained from the patient and family discussed in the report.

The authors state that they have no Conflict of Interest (COI).

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