Achalasia: a rare cause of stridor

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IM - PHYSICAL EXAMINATION

Achalasia: a rare cause of stridor

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Case report

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A 56-year-old woman presented to the emergency department (ED) acutely stridorous. The patient was leaning forward, unable to speak and could not swallow. The patient was, however, able to communicate through gesturing and writing. She confirmed a prior history of untreated achalasia, and had eaten a large meal one hour prior to ED presentation.

Physical examination (PE) revealed a woman in obvious respiratory distress. Vital signs included: temperature 36.4°C, pulse 92 beats per minute, blood pressure 109/56 mmHg, respiratory rate 30 breaths per minute and a room air oxygen saturation of 100%. The head, ears, eyes, nose and throat examinations were unremarkable. The neck examination revealed an audible inspiratory stridor. Palpation of the neck did not identify a mass or thyromegaly, however the patient did have tenderness in the anterior neck. The remainder of the PE was unremarkable.

Radiographs were obtained. A cardiothoracic surgery consultation was obtained. The insertion of a nasogastric tube (NGT) was attempted to decompress the esophageal dilatation. The NGT repeatedly coiled and was unable to be passed. An esophagogastroduodenoscopy (EGD) was then performed. The endoscope, however, could not be advanced due to a large impacted food bolus. After 10 l of saline lavage and forceps removal of food pieces the scope was advanced through the lower esophageal sphincter (LES), which was noted to have increased tone. The patient's stridor resolved, and she remained asymptomatic after the EGD. The patient was discharged to home asymptomatic on hospital day two to follow up for outpatient surgical myotomy.

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Comment: Listening to the stridor

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Stridor is rarely encountered in common clinical practice. Even when matched, of all the diagnosis that ever will be made, achalasia is the most rare. The patient was lucky twice: she had the opportunity to communicate the prior history of untreated achalasia, and she met a listening physician. Indeed, stridor was a marker of an urgent situation, but the patient history was the real guide to a correct diagnostic course.

Stridor is a loud musical sound of definite and constant pitch (usually about 400 Hz) that indicates upper airway obstruction. It is identical to wheezing acoustically in every way except for two characteristics: (1) stridor is confined to inspiration, whereas wheezing is either confined entirely to expiration (30–60% of patients), or occurs during both expiration and inspiration (40–70% of patients) [1, 2]; and (2) stridor is always louder over the neck, whereas wheezing is always louder over the chest [2]. In some patients with upper airway obstruction, stridor does not appear until the patient breathes rapidly through an open mouth. When stridor is present, an airway diameter less than 5 mm can be estimated [3].

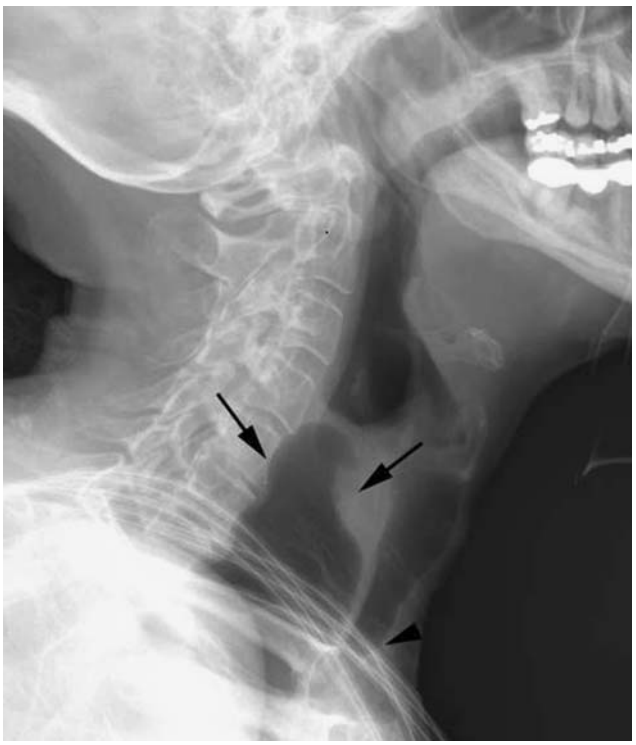

Fig. 1 Lateral neck radiograph demonstrating a widely dilated upper esophagus (*arrows*) and tracheal compression (*arrowhead*)

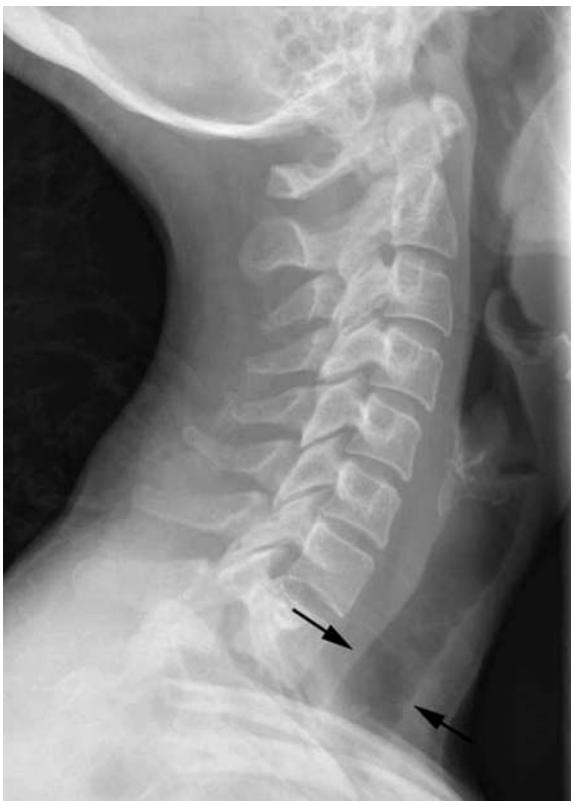

Fig. 3 Comparison view of normal lateral neck radiograph. *Arrows* outline normal tracheal width

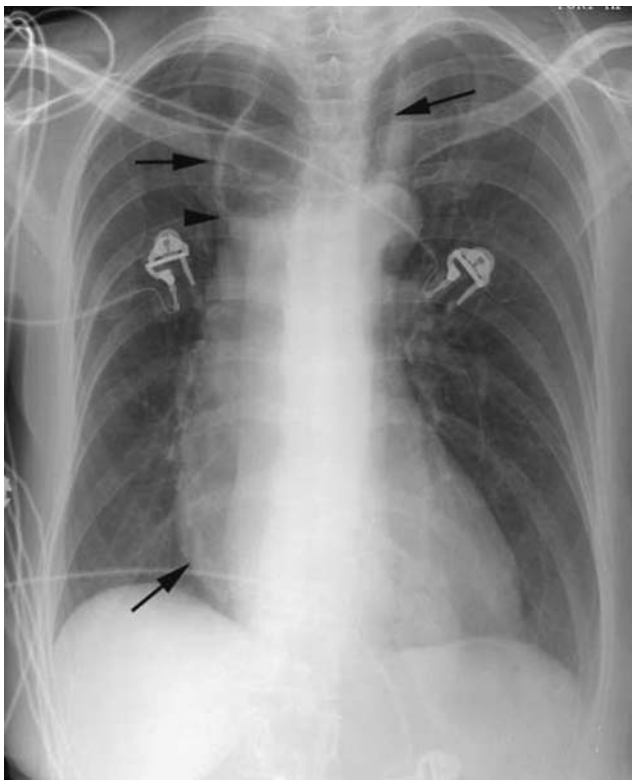

Fig. 2 AP chest radiograph reveals a widely dilated esophagus (*arrows*) and the top of a food bolus impaction (*arrowhead*)

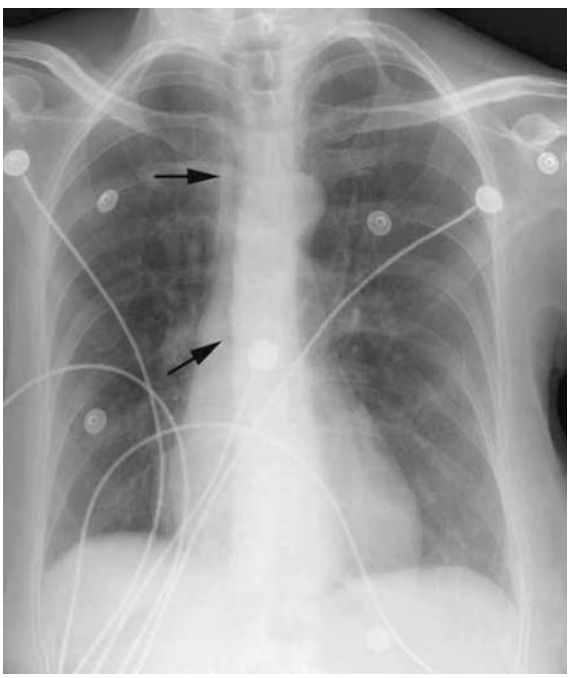

Fig. 4 Post-endoscopic lavage chest radiograph. *Arrows* depict air-filled esophagus

In young children between 6 months and 3 years, croup is one of the most frequent causes of acute respiratory distress with a peak annual incidence in the second year of life of nearly 5%. In children a careful history and physical examination is the best method to confirm the diagnosis, and to rule out potentially serious alternative disorders such as bacterial tracheitis and other rare causes of upper-airway obstruction. Children who have stridor secondary to the presence of a foreign body usually present with a clear history of ingestion. Stridor, indrawing of the sternum, agitation or lethargy, respiratory rate, and pulse rate are also included in different clinical score to assess disease severity [4, 5]. Other methods for objective assessment have been reported but are found to be either impractical, or insensitive [5]. In a retrospective study, children with persistent sternal indrawing at presentation to an ED had a 6% probability for endotrachael intubation, whereas those without sternal and chest-wall indrawing recovered rapidly without any specific treatment [6]. Therefore in children only congenital, chronic, or severe stridor may require direct visualization of the airways with a flexible fiberoptic bronchoscope.

In adults stridor requires more aggressive management than in children, and except for acute allergic reaction, early diagnostic exploration of the upper airway with flexible fiberoptic direct laryngoscopy is usually suggested [7] to exclude the presence of abscess, laryngeal tumor, or aspirated foreign body. Stridor should also warn the clinician of the possible requirement of an artificial airway. However, the starting point for all clinical decision is pretest probability, i.e. the probability of disease (i.e. prevalence) before application of the results of further examination.

A positive history of prolonged tracheal intubation and tracheostomy may suggest a subglottic stenosis, and therefore, a diagnostic exploration of upper airway is indicated. Subglottic stenosis usually causes dyspnoea as the most common presenting symptom rather than stridor. In the presence of stridor, a positive history of achalasia makes high the pretest probability of upper airway obstruction, but it is the oesophagus that needs exploration. Therefore achalasia can be included in a differential diagnosis in all patients presenting with stridor, but oesophagus investigation is mandatory in patients with a past history of the disorder.

The cephalic displacement of the dilated esophageal lumen behind the cricopharyngeus muscle may create a one-way valve that allows air to enter the esophagus on inspiration, but prevents cephalic escape of air [8]. Progressive oesophageal dilatation can cause forward displacement and tracheal compression against the sternum (see Fig. 1), and acute airway distress may result (Figs. 2, 3, 4).

As a consequence the presence of the achalasia history forced a peculiar diagnostic course. A dysfunction of the upper esophageal sphincter (UES), with reduced relaxation with swallowing, made difficult both the escape of air across the UES and nasogastric tube passing. Esophagogastroduodenoscopy was both diagnostic and therapeutic.

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