An Involuntary and Unexpected Treatment of Nutcracker Esophagus

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A 76-year-old woman complained of painful dysphagia and loss of weight. Esophagoscopy results were negative, whereas computed tomography (CT) disclosed a 25-mm mediastinal tumor without a connection to the esophagus. A diagnosis of nutcracker esophagus was made on high-resolution esophageal manometry. Peroral endoscopic esophageal myotomy failed to improve the symptoms. Right video thoracoscopy allowed resection of the tumor, which looked like a neurogenic tumor of the posterior mediastinum that developed from the right vagus nerve. The patient's dysphagia dramatically improved postoperatively. Because the pathologic examination disclosed a benign solitary fibrous tumor of the pleura, we hypothesize that the motility disorder would have been resolved by the unilateral vagotomy.

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The underlying causes of nutcracker esophagus are not well understood, but the condition may be caused by abnormalities in the nerve pathways that control the esophagus. We report a case that suggests the beneficial effect of vagotomy to relieve disabling symptoms.

A 76-year-old woman was admitted to our department for severe spastic and painful dysphagia with a 15-kg loss of weight in the previous year. Her history was unremarkable, except for the curative treatment of breast cancer 3 years before. She had no history of gastroesophageal reflux disease or psychiatric illness.

Esophagoscopy did not reveal any endoluminal abnormality. A diagnosis of nutcracker esophagus was made on the high-resolution esophageal manometry assessment (Fig 1).

Chest computed tomography (CT) found an isolated 25-mm tumor in the right superior and posterior mediastinum located close to the esophagus (Fig 2). Magnetic resonance imaging showed homogeneous enhancement distinct from the esophagus. Endoscopic ultrasonography confirmed the absence of invasion of the external esophageal layers. Transesophageal cytologic examination was not informative. Positron emission tomography scanning disclosed the absence of metabolism.

In the absence of suspected malignancy, priority was given to the esophageal disorder. Esophageal symptoms did not improve despite dietary counseling, medical treatment with nitroglycerin during crisis, and continuous proton pump inhibitors and calcium channel blockers. Two endoscopic procedures were attempted subsequently but unsuccessfully: (1) botulinum toxin injections (100 IU in the last third of the esophagus body with 10 injection sites) and (2) peroral endoscopic myotomy that consisted of a mucosal incision 10 cm above the esophagogastric junction followed by the creation of a submucosal tunnel up to the esophagogastric junction, a myotomy of the circular muscle in between, and then the closure of the mucosal entry with endoscopic clips.

Three months later, surgical exploration of the mediastinal mass was finally performed through a right video thoracoscopic approach. Macroscopically, the tumor looked like a neurogenic tumor of the posterior mediastinum, with the lower pole located just above the level of the azygos vein arch and the upper pole over the thoracic inlet, without any connection with the esophagus. Complete resection was performed en bloc with a segment of the right vagus nerve located distal from the origin of the right recurrent nerve, from which the tumor was thought to have developed.

The postoperative outcome was marked by the immediate and spectacular improvement of esophageal symptoms, which was maintained at the last follow-up 9 months after the operation. Pathologic examination disclosed an encapsulated and multilobulated 35-mm lesion, with homogeneous content and rare cellularity, mainly constituted of fusiform cells, and a negative mitotic index. The final diagnosis was a benign solitary fibrous tumor originating from the mediastinal pleural layer.

Comment

We have reported a case of severe painful dysphagia with loss of weight caused by a nutcracker esophagus, which totally disappeared after resection of a benign solitary fibrous tumor of the mediastinal pleura. This association has never been observed previously, but it is probably coincidental. Indeed, we hypothesize that the right vagotomy, as a consequence of the en bloc resection of what we thought to be a neurogenic tumor of the posterior mediastinum, had a beneficial impact on the esophageal functional disorder.

Solitary fibrous tumors are rare mesenchymal tumors. They affect a wide range of connective tissues, but mainly the pleura. Solitary fibrous tumors of the pleura are most frequently asymptomatic and thus result in an incidental finding on imaging. When they are present, symptoms are usually nonspecific (ie, cough, chest pain, dyspnea). Some associated paraneoplastic syndromes have also been reported, such as a hypertrophic pulmonary osteoarthropathy in 20% of the cases [1]. More rarely,

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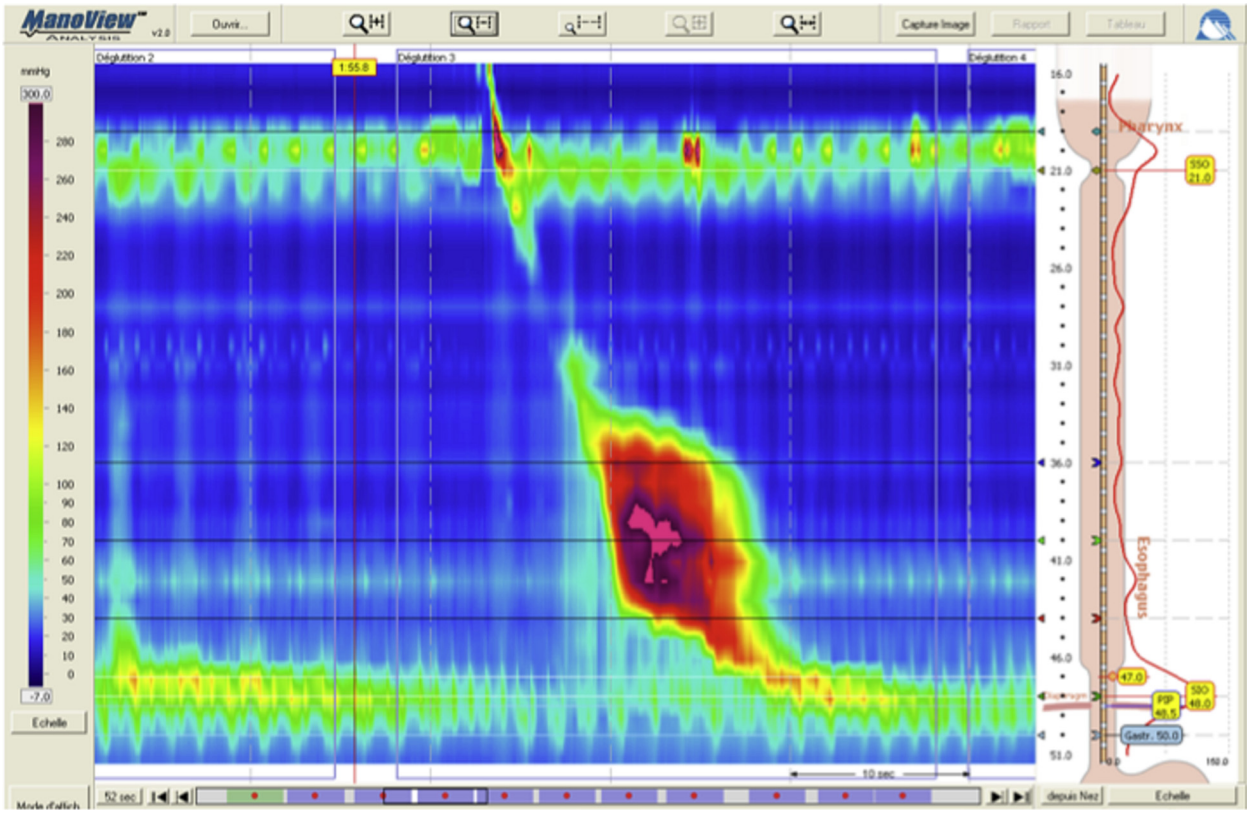

Fig 1. High-resolution esophageal manometry assessment: nutcracker esophagus. Normal pressure into inferior esophageal sphincter, presence of hypertensive peristalsis with mean distal contractile integral greater than 5,000 mm Hg/s/cm, and no swallow distal contractile integral greater than 8000 mm Hg/s/cm.

hypoglycemia resulting from the tumor production of insulin-like growth factor 2, known as Doege-Potter syndrome, is encountered in 2% to 4% of cases [2]. To the best of our knowledge, no paraneoplastic esophageal motility disorders have ever been reported in this setting.

Whatever the clinical context, these tumors should be resected whenever possible, because the diagnosis of benignity is difficult to ascertain. England and colleagues [3] have suggested criteria to diagnose malignancy, including high mitotic activity, high cellularity, necrosis, hemorrhage, and pleomorphism. In addition, de Perrot and associates [4] have proposed a classification based on morphologic features (sessile, pedunculated) and pathologic type (benign, malignant) because of the wide variety of recurrence patterns that justifies a half-yearly radiologic follow-up during the initial 2 years and yearly thereafter.

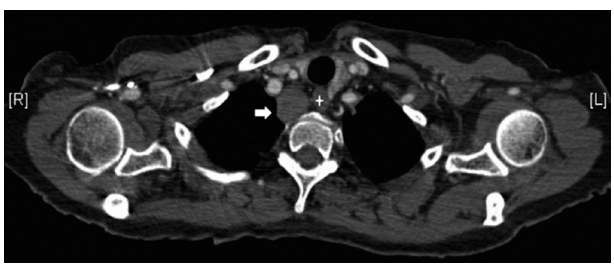

Fig 2. Thoracic computed tomography (CT) with intravascular contrast showing homogeneous mass in right superior mediastinum (white arrow), distinct from esophagus (+) with homogeneous enhancement.

Nutcracker esophagus is an esophageal functional disorder characterized by contractions in the smooth musculature in a normal sequence but at an excessive amplitude or duration. Nutcracker esophagus is 1 of several motility disorders of the esophagus, which include achalasia and diffuse esophageal spasm [5]. Nutcracker esophagus can affect individuals of any age and either sex but, as in our patient, is more common in women and in the sixth and seventh decades of life. The underlying causes of nutcracker esophagus are not well understood, but the condition may be caused by abnormalities in the nerve pathways that control the esophagus. The peripheral neuromuscular control mechanisms involved in peristalsis of the esophageal circular smooth muscle have been an area of intense investigation for many years. The importance of intrinsic neuromuscular mechanisms in the generation of the peristaltic wave is established. Vagal efferent fibers have a crucial role in initiating and modulating esophageal peristalsis [6]. Vagotomy is thus expected to interfere with esophageal motility. Accordingly, pseudoachalasia on an initially

healthy esophagus after bilateral truncal vagotomy has been reported [7]. Unilateral or bilateral lesions of the vagus nerve caused by extensive mediastinal lymph node dissection during lung cancer operations have also been suspected as being responsible for alteration of gastro-esophageal motility [8]. We thus reasonably hypothesize that the inhibitory effects of unilateral vagotomy on esophageal peristalsis might have balanced the intrinsic esophageal hypercontractility in our patient. This observation offers a new insight into the treatment of patients with this rare but disabling condition who do not respond to other therapies.

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