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Periesophageal mediastinal fibromatosis

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Abstract
We report two cases of aggressive fibromatosis of the mediastinum with esophageal involvement in adults. CT showed the posterior mediastinal mass involving the esophagus. In both cases, surgical resection was not feasible. Histopathologic examination showed a fibrous tissue. The patients died two years later.

Key words: Esophageal stenosis—Mediastinal fibromatosis—Fibrous tissue—Mediastinal enlargement—CT of the mediastinum.

Aggressive fibromatosis of the mediastinum is a rare, locally invasive, but nonmetastazing condition. We report two cases and describe the radiographic and computed tomographic appearances.

Case reports

Case 1

A 22-year-old woman presented with painless dysphagia for solids. Two months later, a barium swallow demonstrated regular luminal narrowing of the distal esophagus (Fig. 1A), suggesting peptic stenosis or an intramural esophageal tumor.

Biopsies performed during endoscopy 5 and 6 months after the onset of the dysphagia but 3 and 4 months after the first esophagogram produced negative results. Seven months after the onset of symptoms, a repeat barium study showed the extension of esophageal narrowing (Fig. 1B) to the upper part of the stomach, with features consistent with an intramural mass but with rather slow growth.

Biopsies from the stomach and esophagus showed only mucosal inflammation. Between the two contrast studies, the patient's weight loss was 10 kg in 5 months. The patient eventually had surgery 7 months after the onset of symptoms, just after the second esophagogram.

Tumor involving the gastroesophageal junction and extending to the left lobe of the liver, diaphragm, aortic wall, and the origin of the celiac trunk was found. Although resection was not feasible, celiac and hepatic lymph nodes were removed; frozen section confirmed malignancy.

Thoracic and abdominal computed tomography performed 15 days after surgery showed a vascular tumor involving the entire thickness of the distal esophageal wall (Fig. 1C), the gastroesophageal junction (Fig. 1D), and the upper part of the stomach. This tumor seemed to be adherent to the posterior wall of the liver and to the aorta. A diagnosis of leiomyosarcoma or diffuse esophagogastric leiomyomatosis was suggested.

Pathologic examination of the specimen showed the presence of predominantly fibrous tissue with numerous vessels and inflammatory infiltration. The inflammatory component was polymorphous, but inflammatory cells were predominantly plasma cells. The pathologic diagnosis was aggressive fibromatosis.

The patient died 2 years after the onset of the symptoms from local and regional extension. No postmortem examination was performed.

Case 2

A 61-year-old man presented with an 18-month history of dysphagia. He was otherwise well. A stenosis of the upper esophagus was demonstrated on a barium swallow (Fig.

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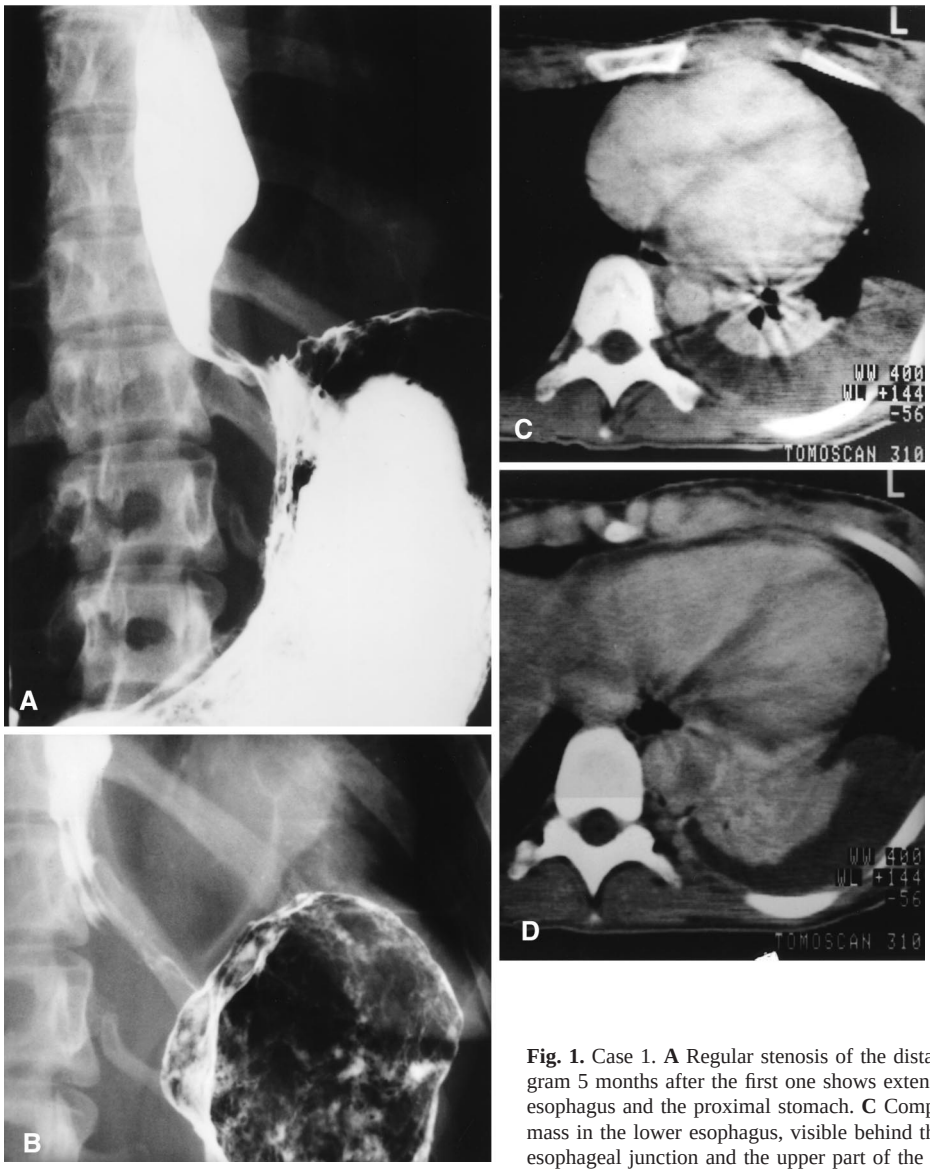

Fig. 1. Case 1. **A** Regular stenosis of the distal esophagus. **B** Second esophagogram 5 months after the first one shows extending luminal stenosis of the lower esophagus and the proximal stomach. **C** Computed tomography shows a dense mass in the lower esophagus, visible behind the heart. **D** Mass in the gastroesophageal junction and the upper part of the stomach.

2A), and endoscopy confirmed marked esophageal stenosis. However, the biopsy showed no endoluminal abnormalities.

The frontal chest radiograph was normal, but tomography (Fig. 2B) and computed tomography showed a large posterior mediastinal mass compressing the posterior part of the trachea. At bronchoscopy, there was displacement of the trachea because of a posterior mass. An exploratory thoracotomy showed a large unresectable posterior mass. Frozen section was suggestive of a fibrosarcoma. However, histopathologic examination showed a fibrous tumor (Fig. 2C) without signs of malignancy. A diagnosis of fibromatosis or fibrosarcoma grade I was suggested.

At the 2-year follow-up, there was enlargement of the upper mediastinum (Fig. 3). The patient started suffering from increasing dyspnea. He died after a bronchoscopy 2 years 2 months after the disease had been discovered. No postmortem examination was performed.

Discussion

Fibromatosis usually presents as large, infiltrative, fibrous soft tissue masses composed of highly differentiated fibroblasts cells, i.e., with intermediate behavior between a benign fibrous lesion and fibrosarcoma [1]. These tumors have a tendency to infiltrate surrounding tissue and to form healed circumscribed tumors. Although fibromatosis tends to recur locally, it does not metastasize.

Idiopathic mediastinal periesophageal fibromatosis is rare and is characterized by the presence of plasmocytic

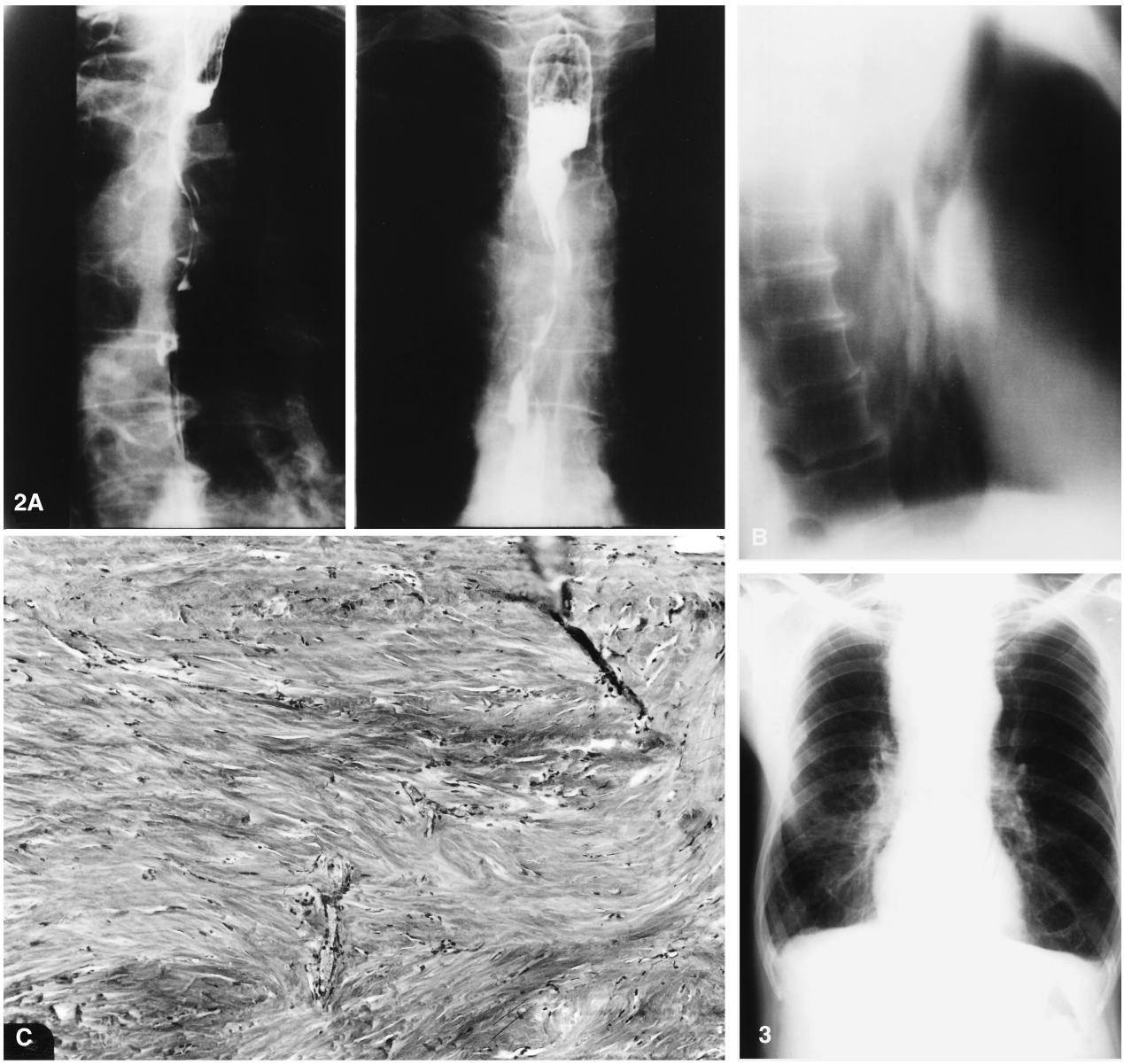

Fig. 2. Case 2. **A** Frontal and oblique view of the esophagus shows extended and marked stenosis of the esophagus with proximal dilatation from T2 to T6. **B** Lateral laminogram of the mediastinum shows posterior mediastinal mass compressing the trachea. **C** Fibrous tumor without signs of malignancy.

Fig. 3. Frontal view shows enlargement of the upper mediastinum.

cell infiltrates and the absence of any sign of malignancy. Most patients present with dysphagia. A barium swallow may show a regular or eccentric stenosis without dilatation of the proximal esophagus, and differentiating between an extrinsic and intramural lesion is difficult.

Endoscopy usually shows marked, regular stenosis with normal mucosa and without an intraluminal mass. The diagnosis of esophageal carcinoma may be excluded based on macroscopic features.

Computed tomography and magnetic resonance imaging may be useful in assessment. Computed tomography [2, 3, 4] typically demonstrates a periesophageal mass and allows differentiation between mediastinal fibrosis and other lipomatous, vascular or cystic lesions and allows proper evaluation of the extension of this lesion, sometimes to the chest wall and intraspinal [5].

Magnetic resonance imaging [6] shows good characterization of lesions and excellent soft tissue contrast. The tumor is iso-intense relative to muscle on T1-weighted images and enhances after intravenous administration of gadolinium. However, the features are nonspecific, and histologic examination is mandatory.

Macroscopically, a fibrous mass with some extension to the mediastinal structures may be seen. The histologic features are those of a nonspecific inflammatory infiltrate, with mainly lymphoplasmocytic cells. The tumor has overall high cellularity but no cellular atypia or detectable mitoses.

The differential diagnosis for such a stenosis of the esophagus includes peptic esophagitis, an infiltrative neoplastic lesion, and diffuse leiomyomatosis [7–10]. With regard to extrinsic lesions, a differential diagnosis includes malignant mediastinitis, often described in the context of breast cancer after a mean delay of 7 years. Differential diagnosis also include stroma reactions often seen secondary to reticulosarcoma, lymphoma, and moderately differentiated fibrosarcoma of the mediastinum. Possible etiology of mediastinal fibrosis includes sarcoidosis, silicosis, histoplasmosis, autoimmune disease, diffuse lupus erythematosus, and iatrogenic fibrosis (radiotherapy, methysergide).

It is noteworthy that once these etiologies have been ruled out, one must consider idiopathic mediastinal fibrosis; this is the most frequent instance, representing about 70% of the cases.

Combined mediastinal and retroperitoneal fibrosis has been reported [11], but there was no retroperitoneal abnormality in our case. A fatal case of aggressive mediastinal and retroperitoneal fibromatosis in a 24-year-old man has been reported [3].

Extensive surgical resection is the only possible curative treatment [1], but it is rarely possible due to the extent of the lesion. If surgical resection cannot be performed, gastrostomy is necessary.

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