

Letters to the Editor

Rare esophageal tumors of mesenchymal origin

Key words: Sarcoma. Mesenchymal tumor. Esophagus.

Dear Editor,

The esophagus is one of the organs of the digestive tract where tumors are very aggressive. They occur in smokers and/or drinkers with clinical dysphagia especially for solids. Most are malignant epithelial adenocarcinoma being the most frequent if located in the distal esophagus and squamous cell carcinoma when they are built in proximal region. Less frequent are tumors of mesenchymal origin.

Case report

We present a 77-year-old male, smoker of 50 packs/year, with chronic obstructive pulmonary disease (COPD) and atrial fibrillation. He was admitted to the emergency because of progressive dysphagia for solids of three months duration and, in the last month, with liquid weight loss and anorexia.

Endoscopy detected an exophytic neof ormation in the middle third of the esophagus causing six centimeters partial stenosis of the lumen. The scan showed an esophageal tumor without local or distant spread. Bronchoscopy was negative for malignancy, thus ruling invasion of the respiratory tree.

The microscopic study showed a mesenchymal spindle cell proliferation arranged in bundles, with increased vascularity and high mitotic index with abundant necrosis (Fig. 1 A and B).

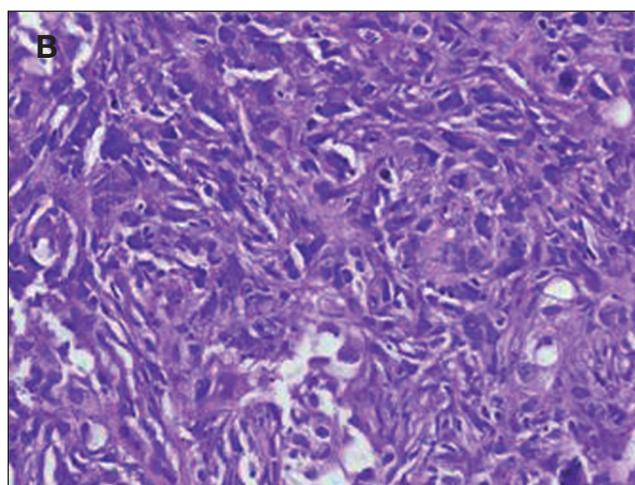
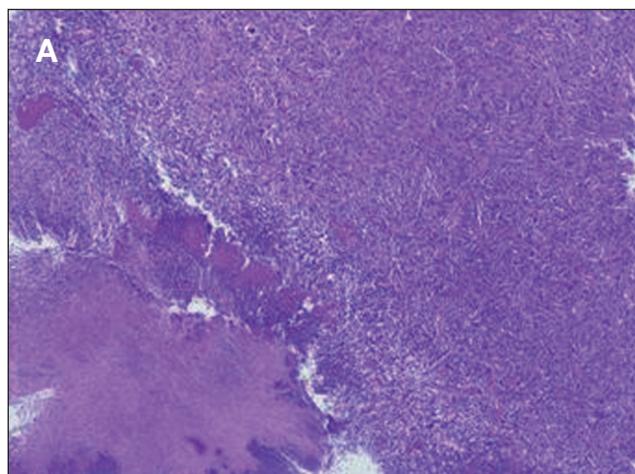


Fig. 1. A. Mesenchymal cells with atypical mitosis and necrosis forming multiple beams stained with hematoxylin and eosin. B. Shows more than one nucleolus indicating loss of normal architecture.

Table I. Differential diagnosis of esophageal tumors according to immunohistochemistry

Tumor	CD117	CD 34	Smooth muscle actin	Protein S100	Desmin	Cytokeratins AE1/AE3, CAM5, 2 and 5/6
GIST	+ > 95%	+ 60-70%	+/- 30-40%	- (5% +)	Rare	-
Leiomyoma	-	+ 10-15%	+	-	+	-
Leiomyosarcoma	-	-	+	-	+	-
Schwannoma	-	-	-	+	-	-
Carcinosarcoma	-	-	-	-	-	+
Sarcoma	-	-	-	-	-	-

Immunohistochemical analysis was negative for desmin, S-100 protein, c-kit and cytokeratin, and C 117 for cytokeratins AE1/AE3, CAM5, 2 and 5/6.

Therefore, we concluded that our patient had a malignant mesenchymal tumor consistent with a poorly differentiated sarcoma. Was assessed for surgical and oncology, dismissing them for impairment of general condition, with stent placement.

Discussion

Sarcomas of the esophagus are very rare, less than 2% of malignant mesenchymal tumors in this location, being leiomyosarcoma and stromal tumors the most frequent (1,2).

Patients are usually males and present with dysphagia. Sometimes, patients complain of respiratory symptoms by invasion of the tracheobronchial tree (3).

Differential diagnosis must be made with B-cell lymphomas, sarcomas (leiomyosarcomas or carcinosarcomas) and metastatic breast carcinoma or melanoma, as well as gastrointestinal stromal tumors, leiomyomas, hamartomas, hemangiomas and granular cell tumors or schwannomas (4,5) (Table I).

The present case was negative for all immunohistochemical markers and fits in the diagnosis of poorly differentiated sarcoma.

Four factors must be taken into account when planning treatment: the clinic, the behavior of the lesion, the patient's condition, and the risk that the surgery is performed.

The treatment of choice is surgery once excluded the presence of local and distant invasion (6-8). However, because few studies have been published and little casuistry has been reported no treatment is protocolized. Attempts have been done to treat advanced

disease with chemotherapy and radiotherapy, but with little success. Therefore, most patients benefit from the placement of a palliative esophageal stent for dysphagia as in our case.

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