Malignant solitary fibrous tumor of the esophagus

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Abstract

Solitary fibrous tumor (SFT) of the esophagus has been very rarely reported in literature. Herein, we report a case of a successful surgically treated malignant solitary fibrous tumor of the esophagus. A 36-year-old woman was admitted at our hospital with an erroneous ultrasound-based diagnosis of cervico-mediastinal goiter. Surprisingly, the preoperative diagnostic work-up, including a computed tomographic chest scan, endoscopy, and endoscopic ultrasonography, revealed a pedunculated intraluminal mass in the esophagus. The tumor was radically removed through left antero-lateral cervicotomy. Pathologic and immunohistochemical examination was concluded for a malignant SFT, a rare variant not previously described in relation to the esophagus. The patient underwent adjuvant radiotherapy and is alive with no signs of tumor recurrence 32 months after surgery.

Keywords: Solitary fibrous tumor; CD34; Esophageal sarcomas

1. Introduction

The solitary fibrous tumor (SFT) is an uncommon mesenchymal tumor first described by Klemperer and Rabin [1] with approximately 800 cases reported in the literature between 1931 and 2002 [2]. It usually originates from the visceral pleura (85—90% of SFT cases), but in recent years, this type of neoplasm has also been reported to occur at numerous extrathoracic sites. In particular, only two cases of benign esophageal SFT have been reported in modern literature [3,4]. Differential diagnosis with other sarcomas is based on pathological examination, especially on the immunohistochemical findings. Surgical excision represents the cornerstone of treatment in SFTs but long-term follow-up is mandatory because of the possibility of late recurrence of these slow-growing tumors. This is the first reported case of malignant SFT of the esophagus.

Herein, we describe its rare presentation, and its radiological and pathological features following excision.

2. Case summary

A 36-year-old woman with a 3-month history of dry cough, dysphagia, and slightly increasing dysphonia was admitted at our hospital with ultrasound (US)-based diagnosis of cervico-mediastinal goiter. All of the laboratory data were within normal limits. Physical examination revealed an elastic mobile mass in the median cervical region.

Contrary to the previous US-based diagnosis, computed tomography (CT) scan showed the presence of a well-circumscribed mass in the upper posterior mediastinum (5.5 cm × 7.5 cm in diameter) with displacement of the trachea and the brachiocefalic vessels. No clear signs of infiltration of the surrounding structures were evident (Fig. 1(A) and (B)).

An endoscopic examination revealed a voluminous mass arising from the esophageal mucosa. Biopsy was undertaken and histology showed a pattern of a malignant neoplasm of mesenchymal origin. A surgical approach was indicated and the patient underwent a left antero-lateral cervicotomy; after hemithyroidectomy (left), the cervical esophagus was prepared and incised. A whitish encapsulated mass (about 7 cm × 5 cm) arising from esophageal mucosa was detected and removed. The incision on the esophageal wall was closed with two-layered sutures and a cervical drain was left in situ up to the 5th postoperative day (after barium swallow radiological check). The patient was discharged on the following day (6th). The tumor macroscopically appeared as a whitish well-circumscribed, elastic, and firm mass (78 mm × 45 mm × 41 mm) with a gray-tan whorled cut surface (Fig. 2(A)).

Histologically, a patternless proliferation of spindle cells, in a richly collagenous matrix with branching capillary channels, large-gaping sinusoidal spaces (‘staghorn’ configuration) was
evidenced thus meeting coherently the definition of ‘heman-
giopericytoma-like vascular pattern’ (Fig. 2(B)). Surgical
borders were disease-free (R0) but neoplastic cells have been
detected 0.5 cm from the cut margins. More than five atypical
mitoses × 10 high power field (HPF) were found. Immunohis-
tochemically, the neoplastic cells showed cytoplasmic immu-
noreactivity to bcl-2 (Fig. 2(C)) and vimentin, and focally to
CD34 (Fig. 2(D)) and CD99, but were consistently negative for
CAM 5.2, AE1/AE3, Desmin, Melan-A, CD 57, and S-100 protein.
Definitive pathology concluded for a malignant SFT. The
patient underwent adjuvant radiotherapy (total dos-
e = 50.4 Gy). Thirty-two months postoperatively, the patient
is alive with no signs of tumor recurrence.

3. Discussion

Because of the variability of pathological morphology
(different patterns or ‘patternless’) the diagnosis of SFT,
above all, in an unusual site such as in the case reported
herein, is particularly difficult.
These tumors are usually observed in middle-aged adults between 20 and 70 years (median 50 years) with no sex predilection [4]. Although SFTs are generally indolent neoplasms which may be cured with complete surgical resection, malignant cases have been reported in 10–15% of thoracic SFTs [5]. Macroscopically, SFT has been described as oval-shaped, well-circumscribed, or pseudo-encapsulated masses with gray to yellowish cut surfaces and firm in consistency. Diagnosis of malignancy was established on the basis of criteria suggested by England et al. [6]: the presence of high cellularity, cellular pleomorphism, necrosis, and high mitotic count (≥4/10 HPF). Microscopically, the tumor is composed of cytologically scattered spindle cells set in a collagenous matrix with typical keloid-like hyalinization and arranged in a ‘patternless’ manner or in a variety of patterns including high-performance computing (HPC)-like vascular pattern and storiform.

The diagnosis of SFTs is based on this characteristic microscopic appearance in combination with immunohistochemical studies. Typically, strong and diffuse positivity for CD34 is currently regarded as the characteristic and a key finding of SFT. Immunohistochemical studies have also shown positive staining of the spindle cells to vimentin, associated with negative results for cytokeratin, s-100 protein, desmin, α-SMA, and CD31 [7].

Despite these histological findings, SFT behavior remains unpredictable and long-term follow-up (wait and watch) is mandatory because of the possibility of late recurrence. As described in the pertinent literature, some tumors, with atypical or malignant histological features, did not behave aggressively and other tumors, without any atypical histological features, developed local recurrence or evolved with the appearance of distant metastases [8].

Furthermore, the number of reported cases of malignant SFT is too small to offer an educated overview. Recurrences are inconsistently reported and no established treatment modality has been agreed to. Therefore, to date, surgical excision represents the cornerstone in SFT treatment. In our case, an adjuvant radiotherapy treatment was performed taking into consideration the relatively young age of the patient, the histological malignant features, and the appearance of surgical borders.

In conclusion, SFTs of the esophagus are a very rare entity with unpredictable behavior. Precise diagnosis is usually based on the correct interpretation of specific pathologic and immunohistochemical features. Surgical excision is mandatory. To the best of our knowledge, this is the first case of malignant SFT of the esophagus.

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References