Case report

VATS resection of an oesophageal leiomyoma in a patient with neurofibromatosis Recklinghausen

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Abstract

A series of reports in the literature suggest an association of neurofibromatosis Recklinghausen with intestinal tumors as carcinoids, leiomyomas and leiomyosarcomas. We present a case of a 23-year-old man with severe cutaneous manifestation of neurofibromatosis. Dysphagia was the main symptom. CT scan suggested the diagnosis of an oesophageal leiomyoma. The oesophageal muscle layers were split and the tumor was enucleated by video assisted thoracoscopic surgery (VATS). The postoperative course was uneventful. The patient was drinking liquids from day 1 and was eating a normal diet from day 3 postoperatively. He was dismissed from the hospital on the 4th postoperative day. We conclude that in patients with neurofibromatosis and oesophageal symptoms an intestinal manifestation of the disease in the oesophagus has to be considered and that VATS resection of intramural and extrinsic oesophageal leiomyomas is the treatment of choice. © 1997 Elsevier Science B.V.

Keywords: Neurofibromatosis; Oesophageal leiomyoma; Video assisted thoracoscopic surgery

1. Introduction

The association of neurofibromatosis Recklinghausen with intestinal tumors as carcinoids, leiomyomas and leiomyosarcomas is documented in the literature in a number of case reports [1–4] and seems to be more than casual. The etiology of this association is unknown. At least 15 patients with neurofibromatosis and intestinal leiomyomas have been reported in the literature [3,6]. Only one case of an oesophageal neurofibroma is well documented [5]. Even neurofibromatosis is associated with intestinal leiomyomas no previous reports of an oesophageal leiomyoma in a patient with neurofibromatosis is described in the literature. Oesophageal leiomyomas can be enucleated endoscopically by videoassisted thoracoscopic surgery (VATS). The technique of resection of this benign oesophageal tumor is presented.

2. Case report

A 23-year-old man with multiple cutaneous manifestations of neurofibromatosis presented with left sided chest pain and dysphagia for solid food since 6 weeks, recurrent vomiting, but stable body weight. Routine laboratory, clinical findings and abdominal sonography were normal. X-ray examination after barium swallow demonstrated a stenosis in the middle third of the oesophagus (Fig. 1a). Endoscopic and endosonographic evaluation confirmed a mid-oesophageal obstructive tumor (3 × 1 × 5 cm) suspicious for leiomyoma. The mucosa did not show any signs of ulceration or infiltration of the tumor.
The leiomyoma was enucleated using video-assisted thoracoscopic surgery (Fig. 2a, b). The postoperative course was uneventful. The chest tubes were removed at the 1st and 3rd postoperative day, respectively. The patient was drinking liquids from day 1 and was eating a normal diet from day 3 postoperatively. He was dismissed from the hospital at the 4th postoperative day. A radiologic control with barium swallow was done 5 days after discharge (Fig. 1b).

3. Endoscopic technique

The patient was intubated with a double lumen endotracheal tube and positioned in the left lateral position. Four open ports (3 x 1 cm and 1 x 5 mm) were placed in the fourth, fifth and sixth intercostal space as depicted in Fig. 2a). The camera was introduced in the median axillary line. The lung was retracted anteriorly and the mediastinal pleura on the oesophagus was opened in cranio caudal direction distal to the azygos vein. The main portion of the tumor was located directly below the vena azygos. Therefore the pleura proximal to the azygos vein was opened and the vessel divided with the vascular stapler (Endo GIA, 30 mm auto-suture). The oesophagus was isolated carefully and encircled with mersilene tape.

The myotomy of the oesophageal muscular layer was performed longitudinally. The extramucosal leiomyoma was enucleated stepwise by blunt dissection. The tumor (3 x 2 x 5 cm) was extracted in an endo-bag through the slightly enlarged anterior incision. The muscular layer of the oesophagus was closed with four single stitches (Dexon 3-0) using extracorporeal knotting technique. Two chest tubes were inserted and the incisions closed.
4. Macroscopic and histologic findings

The tumor measured $3 \times 2 \times 5$ cm and was encapsulated. The cut surface was yellowish-white in color and revealed a whirled appearance.

Histologically the tumor was composed of spindle-shaped cells, arranged in interlacing bundles. The nuclei were generally elongated and showed no significant pleomorphism. Mitoses were very rare. Immunohistochemically the tumor cells stained positive for antibodies to Desmin and $\alpha$-Actin (Fig. 3), markers of smooth muscle cells, and negative for neurofilament protein and cytokeratin.

5. Discussion

Tumors derived from smooth muscle cells count for only 1% of gastrointestinal neoplasias [7]. These are divided in leiomyoma and leiomyosarcoma. Leiomyoma is the most common benign gastrointestinal tumor and occurs in the entire gastrointestinal tract. Autopsy studies demonstrated an incidence of 0.07–0.4% [8]. The stomach (66%) is the most frequent location followed by small bowel (24%), oesophagus (7%) and large bowel (3%) [7,9]. These tumors may cause gastrointestinal bleeding [1] but the rate of malignant degeneration is smaller in the esophagus than in other localizations in the GI tract. A number of case reports indicate an association of neurofibromatosis with the occurrence of gastrointestinal tumors which include leiomyoma [1–4], leiomyosarcoma [3], and neuroendocrine tumors (carcinoids) [10]. However, this report seems to demonstrate the first patient with neurofibromatosis Recklinghausen and an oesophageal leiomyoma. The patient presented with chest pain and dysphagia, which are the most typical symptoms of oesophageal leiomyoma. Further symptoms may include cough, gastrointestinal bleeding and weight loss.

A thoracoscopic approach to enucleate the leiomyoma was chosen as described by Everitt et al. [11] and
Bardini et al. [12]. In contrast to previous reports [11] only four ports were used. The extramucosal resection was easily performed and oesophagoscopy with air insufflation to confirm an intact mucosal layer was not necessary. The oesophageal muscular layer was carefully closed because of the reported development of pseudodiverticles after enucleation of leiomyomas [12,13]. The barium swallow 9 days postoperatively showed normal oesophageal passage.

We conclude that in patients with neurofibromatosis and oesophageal symptoms an intestinal manifestation of the disease in the oesophagus has to be considered. Additionally, this case demonstrates that enucleation of an intramural or extrinsic oesophageal leiomyoma is an ideal indication for VATS as this benign tumor is easily exposed and safely resected by endoscopy.

References