Concurrent benign schwannoma of oesophagus and posterior mediastinum

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Abstract

A 52-year-old female with recent onset dysphagia and haematemesis was found to have an intramural tumour of the oesophagus. A separate tumour in the posterior mediastinum was also detected. Both the tumours were immunohistochemically and histomorphologically compatible with benign schwannoma. Oesophageal schwannoma is extremely rare and its association with a concurrent schwannoma in posterior mediastinum is not reported earlier in the literature.

Keywords: Schwannoma; Oesophagus; Posterior mediastinum; Dysphagia

1. Introduction

Schwannomas are tumours of the peripheral nervous system composed of neoplastic Schwann cells. The majority of them follow a benign clinical course. Histologically, these tumours are encapsulated, highly vascular, and composed of a homogenous pattern of biphasic fusiform-shaped cells that may have a palisaded appearance. They remain asymptomatic until they attain a significantly large size. Herein, we present a middle aged female with dysphagia due to oesophageal schwannoma who also had a concurrent posterior mediastinal schwannoma.

2. Case report

A 52-year-old female presented with dysphagia to solid food of 15 days duration. She had an episode of haematemesis during this symptomatic period. Upper gastrointestinal endoscopy was done to evaluate the cause of dysphagia and haematemesis. It showed a large nodular lesion at 30 cm from incisors, occupying entire lumen of the oesophagus (Fig. 1e). Chest X-ray showed a right perihilar mass (Fig. 2a). Computed tomography of the chest showed a hypo dense soft tissue mass in the oesophageal lumen with proximal dilatation. It also showed a similar hypo dense soft tissue mass in the right paravertebral gutter without any intraspinal extension (Fig. 2b). Computed tomography guided tru-cut biopsy from the posterior mediastinal mass and fine needle aspirate from the oesophageal mass confirmed them as benign spindle cell lesions. A provisional diagnosis of oesophageal leiomyoma with a separate neurogenic tumour in the posterior mediastinum was made.

Physical examination, preoperative laboratory investigations and pulmonary function test of the patient were normal. Transthoracic approach to both the tumours was planned with selective one lung ventilation using a leftsided double lumen endotracheal tube. The right chest was opened through a right posterolateral thoracotomy at the fifth intercostal space. After division of inferior pulmonary ligament and mediastinal pleura posterior to lung hilum, two discrete tumours were identified (Fig. 1a). The posterior mediastinal mass (size: 9 x 6 cm) was mobilized first and resected with its intact capsule. Intra-operative frozen section biopsy from the mass was reported as benign spindle cell lesion. The oesophagus was mobilized from the neck to diaphragmatic hiatus and the tumour was assessed for possibility of enucleation. The tumour was ~6 x 5 cm involving the oesophageal wall and not amenable to enucleation. Trans-thoracic oesophagectomy with gastric pull-up and cervical oesophage-gastric anastomosis was done. The cut section of the oesophagus showed a globular tumour projecting into its lumen (Fig. 1d).

Histopathological examination of the tumours showed a spindle cell lesion with mild to moderate pleomorphism without mitotic figures. Immunohistochemically the tumours were positive for S-100 protein and negative for CD 117, CD 34 and smooth muscle actin. The tumours were histomorphologically (Fig. 1b) and immunohistochemically (Fig. 1c) compatible with benign schwannoma. Postoperative recovery was uneventful and she was discharged on postoperative day 10.
Oesophageal schwannoma is uncommon and difficult to diagnose on preoperative investigations. All the reports in literature have mentioned about this difficulty due to its similarity in clinical presentation, radiological and endoscopic appearance with gastrointestinal smooth muscle tumour of the oesophagus [1–3]. The majority of the reported oesophageal schwannomas were seen in women [1]. Oesophageal schwannoma commonly presents with dysphagia. It is also known to present with dyspnoea and stridor due to compression of trachea [4, 5]. It can present with haematemeses also, as in our case. Computed tomography and oesophagoscopy findings are similar to any submucosal lesion of the oesophagus. Final diagnosis is possible on positive immunohistochemical staining for S-100 protein and negative staining for smooth muscle markers like CD 117 (c-kit protein), CD 34 and smooth muscle actin [6, 7]. In our patient, the upper gastrointestinal tract endoscopy done to identify the cause of haematemeses did not show any mucosal breach in the oesophagus or stomach. Possibly, the superficial epithelial breach responsible for haematemeses had healed by the time the patient presented to our clinic. The preferred treatment for benign oesophageal schwannoma is enucleation of the tumour with preservation of native oesophagus [2, 3]. The majority of the patients with benign oesophageal lesion do not require esophagectomy. However, for selected large size tumours, esophagectomy may be needed and has been reported [8, 9]. The tumours in the posterior mediastinum are most commonly neurogenic and originate from the neural crest during the development of peripheral nerves. Most of the tumours remain asymptomatic and detected incidentally on chest radiograph or computed tomography. Surgical excision is recommended due to uncertainty of preoperative diagnosis and possibility of malignancy [10].

The authors would like to highlight the diagnostic dilemma due to presence of two separate tumours on preoperative investigations. We thought of the possibility of presence of a leiomyoma of the oesophagus with large extraluminal component. However, the radiologist, after careful examination of multiple sections at different levels, was categorical about the presence of two separate tumours. The presence of two separate tumours in the oesophagus and paravertebral area was confirmed at the operation. The other possibility considered was of malignant gastrointestinal smooth muscle tumour of the oesophagus with a metastatic growth in the posterior mediastinum. Intraoperative frozen section biopsy of posterior mediastinal tumour ruled out malignancy. We acknowledge that enucleation is the preferred treatment for benign encapsulated tumour of the oesophagus. The tumour in our patient was also assessed for enucleation through a longitudinal incision in the muscularis layer of the oesophagus over the tumour. Enucleation of the lesion was tried through blunt and sharp dissection but the procedure started to cause tear in oesophageal mucosa. Therefore, it was abandoned and oesophagectomy with gastric pull-up was performed. The excised specimen was examined immediately after surgery and the tumour was found to be adherent to mucosa of the oesophagus.

3. Discussion

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