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CASE REPORT

Concomitant Esophageal and Pulmonary Neurofibroma Independent of Neurofibromatosis Type 1: A Rare Case Report

Manabendra Biswas[1], Mosharraf Hossain [2], Delwar Hossain [3], Asit Baran Adhikary [4], Md. Mushfiqur Rahman [5]

Abstract

Concomitant occurrence of esophageal and pulmonary neurofibroma without neurofibromatosis type 1 is exceedingly rare. We are reporting such a case; a 25 year old woman was under evaluation for her painless progressive dysphagia within a short period with history of intermittent fever. Barium esophagogram and esophgaoscopy revealed narrowing of mid and part of lower thoracic esophagus. A CT scan of chest with oral contrast delineated sub-segmental consolidation at posterior basal segment of lower lobe of right lung as well as a long segment circumferential wall thickening in 82.2 mm length of thoracic esophagus extending upto16.4 cm proximal to gastroesophageal junction with proximal dilatation. Patient underwent surgical management and diagnosis was confirmed by histopathology. Patient was discharged home thereafter with marked symptomatic improvement.

Key Words: *Esophageal neurofibroma, Pulmonary neurofibroma.*

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Introduction:

A neurofibroma is a benign tumour originating from nonmyelinating-type Schwann cells in nerve sheath in the peripheral nervous system. In most of the cases (about 90%) they occur as a isolated tumour and only in 10% cases they occur in persons with neurofibromatosis type 1 (NF1),an autosomal dominant genetically inherited disease[1]. The thoracic cavity contains numerous nervous structures from which neurogenic structures may arise, but esophageal and intrapulmonary neurogenic tumour occur infrequently. Esophageal neurofibroma is a benign tumour accounts for only 0.9% all benign esophageal neoplasm[2]. On the other hand, pulmonary neurofibroma accounts for less than 1% of all lung neoplasm[3]. Therefore, concomitant presence of esophageal and pulmonary neurofibroma without neurofibromatosis type 1 is a extremely rare case to be presented. Herein, we described such a case confirmed by histopthologic examination and was managed surgically as per indication.

Case Report:

A 26 year old woman presented to the Thoracic Surgery department of National Institute of Diseases of the Chest and Hospital (NIDCH), Dhaka with painless, progressive dysphagia from solids to liquids, developed over a period of 4 months with sensation of sticking of food. She had no history of weight loss, appetite loss and symptoms of gastroesophageal reflux. She denied any history of corrosive ingestion, foreign

1. Associate Professor, Department of Thoracic Surgery, NIDCH.
2. Associate Professor, Department of Thoracic Surgery, NIDCH.
3. Assistant Professor, Department of Thoracic Surgery, NIDCH.
4. Professor and Chairman, Department of Cardiac Surgery, BSMMU.
5. Resident, MS (Cardiovascular and Thoracic Surgery), Phase-B, BSMMU.

Correspondence to: Dr. Manabendra Biswas, Associate Professor, Department of Thoracic Surgery, NIDCH. Mobile: 01711074483, E-mail: manabendrathoracic@gmail.com

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body impaction, esophageal instrumentation or chest radiotherapy. She also had fever which occurred intermittently over the preceding 3 months; the fever was high grade, continued, not associated with evening rise of temperature or chill and rigor, subsides with paracetamol and used to recur 2-3 weeks later. She had no associated cough, dyspnea, productive sputum, haemoptysis or chest pain. She is a non smoker, non alcoholic and has no history of tobacco use or betel nut chewing. No family history of neurofibromatosis type-1. General examination and systemic examinations including alimentary system and respiratory system examination were unremarkable including absence of skin lesion that of neurofibromatosis type-1.

Routine laboratory investigations including complete blood count, liver and renal function tests were within normal range. Barium swallow X-ray of esophagus (fig-1) revealed narrowing in the midpart of esophagus with proximal dilatation with normal mucosal patterns. Upper gastroitestinal tract endoscopy disclosed narrowing of esophagus at 30 cm from upper incisor teeth with normal overlying mucosa and scope couldn't pass beyond the narrowing. CT scan of chest with contrast (fig-2) outlined a long segment circumferential wall thickening measuring about 17.6 mm in single wall thickness with luminal narrowing at mid and part of lower thoracic esophagus. Involved esophageal segment is approximately 82.2 mm in length and extending upto 16.4 mm proximal to gastro-esophageal junction. Proximal to this lesion esophagus was found dilated. There was sub segmental consolidation at posterior basal segment of lower lobe of right lung (fig-3). No associated mediastinal lymhadenopathy was present.Because of the refractory symptoms and long segment esophageal lesion, patient was

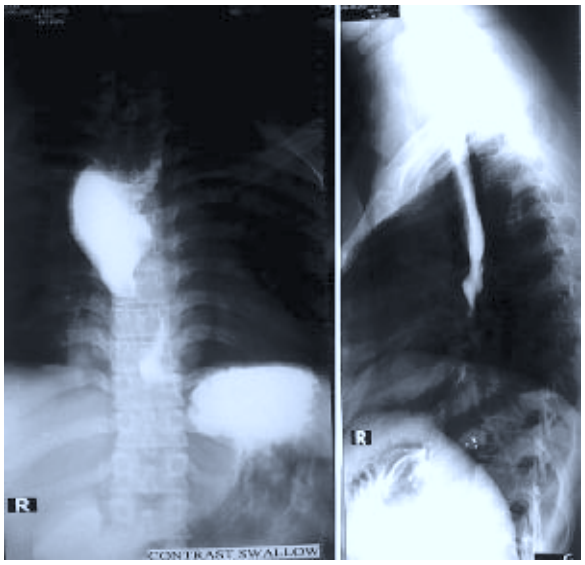

Fig-1: *Barium swallow X-ray of esophagus showing narrowing in the midpart of esophagus with proximal dilatation with normal mucosal patterns.*

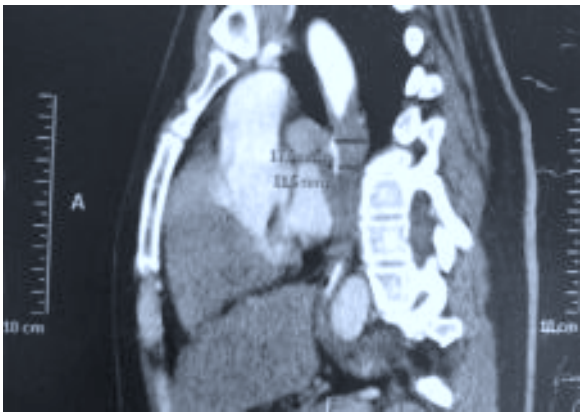

Fig-2: *CT scan of chest showing a long segment circumferential wall thickening with luminal narrowing at mid and part of lower thoracic esophagus.*

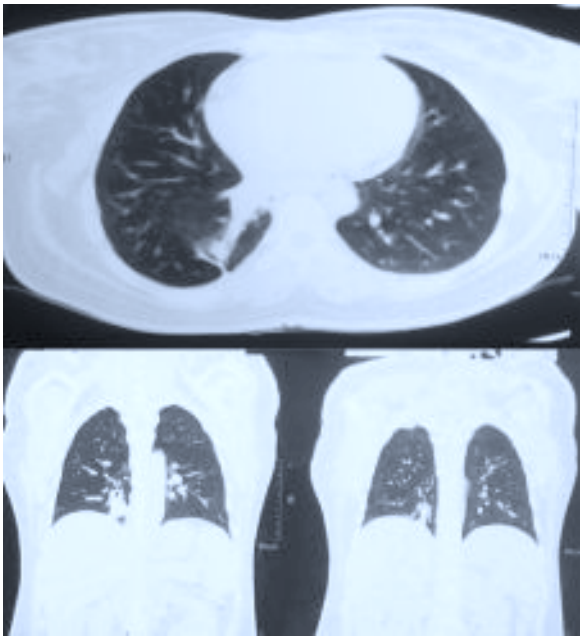

Fig-3: *Sub segmental consolidation at posterior basal segment of lower lobe of right lung*

prepared for two stage esophago-gastrectomy with esophago-gastrostomy. With all aseptic precaution under general anaesthesia abdomen was opened initially with upper midline incision and stomach was mobilized to use as esophageal substitute. Then in the next stage, standard right posterolateral thoracotomy done and chest cavity was accessed. A mass palapated within the lower lobe of right lung which was approximately 3cm x 2cm in size, firm in consistency, therefore right lower lobectomy was done. A mass was also palpated within the middle and part of lower 1/ 3rd of thoracic esophagus which was about 6cm x 2cm size, firm in consistency (fig-4). Esophago-gastrectomy was done containing the mass followed by intrathoracic esophago-

gastrostomy. There were no enlarged mediastinal lymph nodes detected. Both resected specimen, the esophagus and the lower lobe of right lung was sent for histopahology. The hispathology revealed that, macroscopically both the resected specimen of the esophagus and the lower lobe of right lung has a grey-white tumour within, microscopy showed it consists of spindle shaped cells with wavy nuclei, fibroblasts, collagen fibres, neuritis, blood vessels and absence of malignant cells; thereby esophageal neurofibroma and pulmonary neurofibroma was confirmed. The post operative period was uneventful. Post operative contrast swallow X-ray revealed no evidence of leakage from the anastomosis site and no evidence of esophageal narrowing. Post operative CT scan of chest was also satisfactory. Therefore, the patient was discharged on the 10th post operative day with advice including follow up schedule.

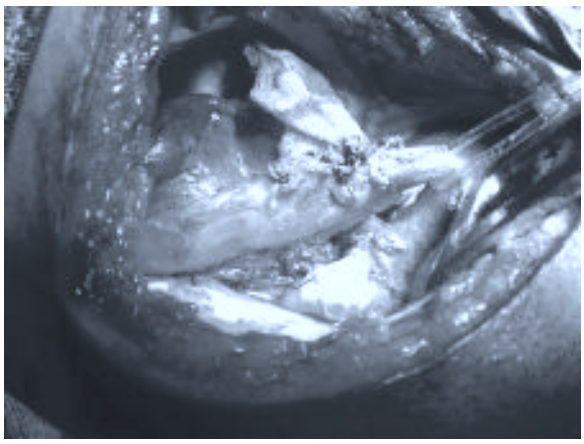

Fig-4: *A mass (Neurofibroma) in the middle and part of lower thoracic esophagus.*

Discussion:

Esophageal tumours are mostly malignant as benign tumors of the esophagus constitute less than 1% of esophageal neoplasms. Nearly two thirds of benign esophageal tumors are leiomyomas whereas neurofibroma are exceedingly rare[4]. Esophageal neurofibromas are of 3 types- localized, diffuse and plexiform. Localized and diffuse neurofibroma are associated with neurofibromatosis type 1 whreas localized neurofibroma usually occurs sporadically as in our patient and arise from Aurbach's plexus and Meissner's submucosal plexus in esophagus[5,6]. Esophageal neurofibroma may remain asymptomatic for years or may present with painless progressive dysphagia within a short period of months like our patient which is also supported by study of others[7,8]. Contrast esophagogram may be done initially to evaluate the patient with dysphagia. CT scan of chest with oral contrast helps to delinate the relationship between the tumour and surrounding structures and also detects mediastinal or extraesophageal pathology. Endoscopic assessment of esophagus is necessary to evaluate the degree of narrowing, condition of the mucosa and to take biopsy in suspicious lesion to rule out malignancy[9]. All the above mentioned tests were done in our case preoperatively except the endoscopy guided biopsy as the esophageal mucosa was healthy in our patient. Surgery is indicated in our patient as the tumour is more than 1cm and the patient is symptomatic[10,11]. Indication for surgery also includes increase in the size of the tumour during follow up. Our patient needed esophageal resection and esophago-gastric anastomosis for refractory symptomps which is also observed in the study of others[12]. Our patient also had pulmonary neurofibroma which may occur either as an endobronchial tumour where trachea being most common site followed by right sided lobar bronchi or may occur as a parenchymal mass like our case[7]. The sporadic pulmonary nerofibroma as in our patient is independent of neurofibromatosis type 1 and arise from bronchial submucosal nerves. A patient with pulmonary neurofibroma may remain asymptomatic similar to our case or may present with cough, dyspnea, wheezing, chest pain and even haemoptysis may occur due to hypervascular nature of the tumour. CT scan of chest, bronchoscopy and guided biopsy followed by

histopathology aid in establish the diagnosis.Depending upon the site and size of the tumour, pulmonary nerofibroma may require surgical resection including wedge resection, segmentectomy, lobectomy besides enucleation of the tumour and bronchoscopic resection[13]. Surgical resection of pulmonary neurofibroma is also indicated where diagnosis is uncertain preoperatively as in our case and also in cases where there is evidence of extrabronchial extension of the tumour or anticipated difficulty with bronchoscopic management including risk of haemorrhage because of hyper vascularity of the tumour[14]. Our patient needed lobectomy which is supported by other studies[13]. Histopathology confirms the diagnosis of pulmonary and esophageal neruofibroma in our patient that comprises spindle shaped cells with wavy nuclei associated with collagen fibrills which is also observed in study of others[2,8,13,15]. The patient had a uneventful intra operative and postoperative period with symptomatic improvement therefore discharged home accordingly.

Conclusion:

The concomitant occurrence of esophageal and pulmonary neurofibroma in our patient purely represent a rare coincidence as they are independent of neurofibromatosis type 1. However, it causes considerable problems preoperatively differentiating from malignant lesion because of their rarity as benign tumour in these organs. Therefore, histopathologic confirmation is done for proper management and follow up plan. Surgical resection usually cures the patients but patients still need to be evaluated during scheduled follow up for recurrence and malignant transformation although rare in sporadic neurofibroma as in this casse.

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