Non-functional paraganglioma of the mediastinum

Niaz Hussain Soomro,[1] Amna Bine Zahid,[2] Aneeqa Ahsan Zafar[3]

Abstract

Paragangliomas are rare tumours which have the ability to secrete neuropeptide and catecholamines. Frequently, the clinical presentation of these tumours is ill defined and some may have no symptoms whatsoever until the tumour is large enough to cause symptoms secondary to local invasion and mass effect, hence making paragangliomas a challenge to diagnose and manage surgically. We report a case of a 55 year old female who presented with shortness of breath and chest pain for 3 years. Further investigations led to the demonstration of a mass along the left side of the mediastinum which was suggestive of a neoplasm. Complete surgical resection of the tumour was achieved and the patient was discharged uneventfully.

Keywords: Paraganglioma, Endocrine tumour, Mediastinal mass.

Introduction

Mediastinal masses maybe benign or malignant tumours that arise within different regions of the mediastinum and are characterized according to the location they originate from; anterior, middle and posterior mediastinum. Radiological investigations like Chest X-ray and CT scan chest with contrast aid in delineating the extent of the tumour and its location.[1] Thymomas and germ cell tumours are most common lesions in anterior mediastinum while lymphomas, mediastinal cysts, and neurogenic neoplasms are the most common primary middle and posterior mediastinal tumours. Treatment involves surgical resection for lesions that appear clinically and radiologically benign while tumours that appear malignant on presentation are usually biopsied first followed by surgery depending on the location and spread of the tumour. Mediastinal paragangliomas are extra adrenal chromaffin cell tumours that account for fewer than 10% of all mediastinal neuroendocrine tumours.[2] They represent only 2% of all catecholamine secreting tumours of the body and 0.3% of all mediastinal tumours.[2] Paragangliomas maybe functional, if they synthesize and secrete catecholamines or non functional, if they synthesize but donot secrete catecholamines.[3] Functional paranganglioma account for 5-15% of all pheochromocytomas. Nonfunctional paragangliomas constitute a very small percentage of these tumours.[4] We report the case of a 55 year old female who was diagnosed with non-functional paraganglioma of the mediastinum.

Case Report

A 55 year old lady presented with shortness of breath and left sided chest pain posteriorly for 3 years. Upon further inquiry, she denied any symptoms of hypertension and headaches, palpitations, sweating, nausea, weight loss, flushing or visual changes. She was afebrile with BP: 110/80mmHg, pulse: 86, RR: 12. General physical examination was unremarkable. No positive findings were noted on examination of the chest. Chest X-ray (Figure-1) revealed a non-homogenous opacity in the left hemithorax at the junction of upper and middle zones along with the mediastinum. These findings were

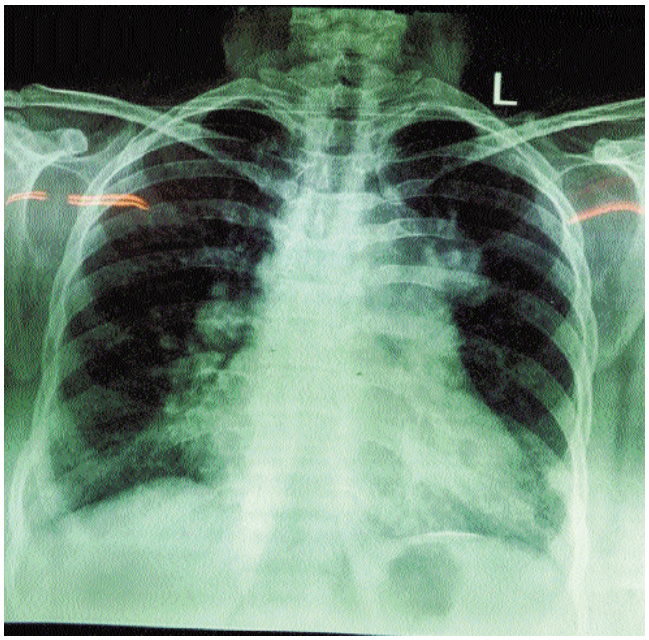

Figure-1: A non-homogenous opacity present on the left side of the chest at the junction of the upper and middle zones along with the mediastinum.

[1]Department of Thoracic Surgery, [2]Intern, Department of Thoracic Surgery, [3]Research Assistant, Ojha Institute of Chest Diseases, Dow University of Health Sciences, Karachi.

Correspondence: Niaz Hussain Soomro. Email: soomroniaz@yahoo.com

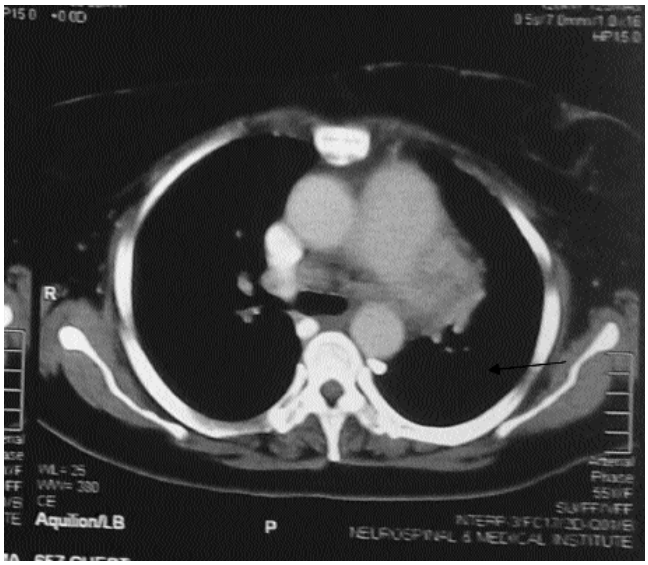

Figure-2: CT-scan chest(mediastinal window) shows a soft tissue density mass(arrow) present along the pulmonary artery and aorta.

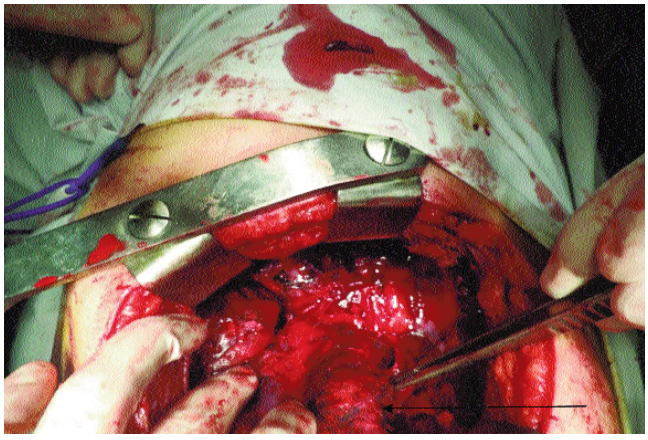

Figure-3: Per operative picture showing left lung retracted medially, with forceps indicating the tumour between the arch of aorta and lung (arrow).

confirmed by CT scan chest (Figure-2). Biopsy of the tumour was performed and histopathology confirmed the diagnosis of paraganglioma. Urinary Vanillylmandelic Acid (VMA) were within normal reference range. Definitive surgery was planned. After pre-operative work up, left sided thoracotomy and excision of the tumour was performed. Per-operatively, a hard mass of about 6cm x 5cm was found in the aorto pulmonary area below the arch of aorta along with the pulmonary artery but not invading the vessels. Tumour was completely excised. Patient's post operative recovery was smooth and she was discharged after 10 days. Her blood pressure was regularly monitored pre, per and post operatively and was found to

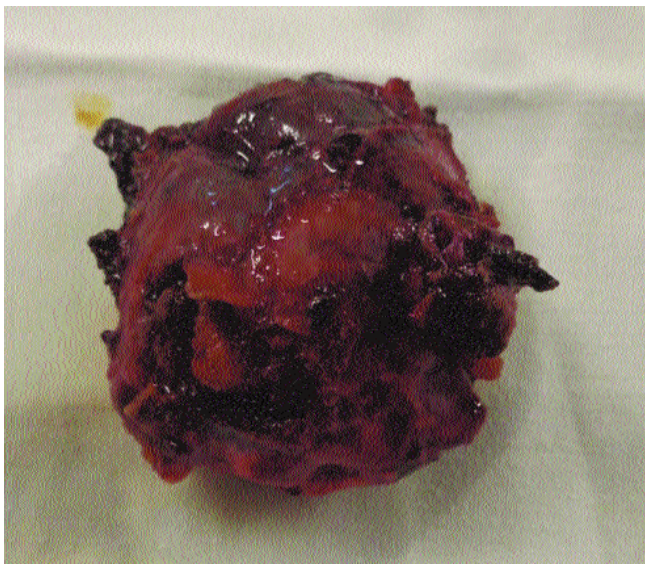

Figure-4: Tumour after excision.

be within normal limits. She is doing well at 6 months follow up. The patient was recently seen on 9 months follow up and she is doing well.

Discussion

The etymology of paraganglioma stems from extra adrenal pheochromocytomas.[5] They are further subcategorized as functional or non-functional based on their ability to synthesize and release catecholamines in the circulation. Mediastinal paragangliomas are hypervascular tumours and many of them invade or firmly adhere to the adjacent mediastinal organs such as heart, great vessels, trachea and the spine.[1] Mediastinal paragangliomas are predominantly concentrated in two locations; the aorticosympathetic paraganglia of the posterior mediastinum or the autonomic ganglia in the superior and middle mediastinum.[6] Aorticopulmonary tumours tend to occur in patients with a mean age of 49 years with no gender preference and only 3% of these secrete catecholamines. The paravertebral paragangliomas, on the other hand, occur in younger people with a mean age of 29 years and almost half of these tumours synthesize catecholamines.[4] Our patient was 55 years old and she presented with a non-functional paraganglioma of the aortopulmonary region. The clinical presentation of paragangliomas ranges from local pressure symptoms, especially in the case of nonfunctional tumours to symptoms related to the systemic effects of secreted cathecholamines and their metabolites in functional tumours. Hypertension seems to be the most common symptom.[7] Non functional paragangliomas maybe discovered incidentally on chest

radiography performed for other reasons. Confirmation of the diagnosis depends upon the functionality of the tumour. Certain immunohistochemical markers can aid in the diagnosis of paragangliomas. Tumours may show positivity for S-100, chrommogranin, synaptophysin, neuron specific enolase and CD-56.[8,9] The tumour in our patient was positive for chrommogranin, synaptophysin and CD-56 and negative for cytokeratin CAM 5.2 Initial assessment of functional chrommafin cell tumours can be done by measuring plasma free and urinary fractionated metanephrines. Urinary VMA was within reference range in our patient. The imaging modalities utilized for paragangliomas include CT, MRI, echocardiography and chest radiography with barium swallow to locate the extent of tumour and relation to surrounding structures.[4] Metaiodobenzylguanidine (MIBG) scans and somatostatin receptor scintigraphy is used for screening for metastatic disease and staging. Preoperative localization is essential in the management of paragangliomas of the thorax as surgical planning and determination of operative approach is heavily dependent upon precise anatomic localization and relationship of the tumour to surrounding structures. Mediastinal paragangliomas behave as low grade indolent neoplasms. Approximately 10 % of cases are associated with metastasis.[2] John et al[10] reported that the most common metastastic sites were local lymph nodes, bone, liver and lung. Complete surgical resection of the tumour, although challenging, renders long term survival rates upto 84% making it the treatment modality of choice.[10]

Conclusion

Paragangliomas are tumours that very rarely arise from the mediastinum. These tumours usually get diagnosed late due to their low incidence and lack of catecholamine secretion. Due to the rarity of these masses, it is necessary to report them in literature so as to create awareness regarding them.

References

1- Milka M, Braham E, Hamrouni R, Zribi H. About a challenging mediastinal tumor. Asian Cardiovasc Thorac Ann 2015; 23: 1125-8
2- Suster S, Moran C. Neuroendocrine tumors of the mediastinum. Am J Clin Pathol 2001; 115(suppl 1): S17-27
3- Earnest LE, Cubilla AL, Woodruff J. Paragangliomas of the head and neck region; A pathologic study of tumors from 71 patients. Hum Pathol J 1979; 10: 191-218
4- Ma L, Mei J, Liu L. Thoracoscopic resection of functional posterior mediastinalparaganglioma;case report. J Thorac Dis 2014; 6: 1861-4.
5- Tiscler AS, Kimura N, Mcnicol A. Pathology of pheochromocytoma and extra adrenal paraganglioma. Ann NY Acad Sci 2006; 1073: 557-70
6- Rakovich G, Ferraro P, Therasse E. Preoperative embolization in the management of a mediastinal paraganglioma. Ann Thoracic surg 2001; 72: 601-3
7- Goldstien RE, O Neil Jr, Holcomb GW 3rd, Morgan WM 3rd, Neblett WW 3rd, Oates JA, et al. Clinical experience over 48 years with pheochromocytoma. Ann Surg 1999; 229: 755-64
8- Pavai Z, Orosz Z, Horváth E, Seres-Sturm L, Jung J. Immunohistochemical features of paragangliomas. J Cell Mol Med 2001; 5: 311-6
9- Kliewer KE, Wen DR, Cancilla PA, Cochran AJ. Paragangliomas: assessment of prognosis by histological, immunochemical and ultrastructural techniques. Hum Pathol 1989; 20: 29-39.
10- Lin MW, Chang YL, Lee YC, Huang PM. Non functionalparaganglioma of the posterior mediastinum. Interact Cardiovasc Thorac Surg 2009; 9: 540-2.