



Case Report

Nonfunctional posterior mediastinal paraganglioma



El Hassane Kabiri*, Massine El Hammoumi, Faycal El Oueriachi, Adil Arsalane

Department of Thoracic Surgery, Mohamed V Military University Hospital, Faculté de Médecine et de Pharmacie,
Université Mohamed V Souissi, Rabat, Morocco

ARTICLE INFO

Article history:

Received 25 October 2012

Received in revised form

7 November 2012

Accepted 4 March 2013

Keywords:

Mediastinal tumor

Paraganglioma

Surgical treatment

ABSTRACT

A 54-year-old man was admitted to our hospital for left chest pain. Imaging studies (chest radiography, computed tomography, and magnetic nuclear resonance imaging) showed a well-defined left paraspinal mass with central necrosis ($6 \times 5 \times 4$ cm), located between T7 and T9. Urine levels of metanephrine and normetanephrine were within normal limits. The patient underwent a complete resection of the tumor through a posterolateral thoracotomy. Perioperative vital signs were normal. The diagnosis of paraganglioma was confirmed histologically. The patient was discharged on the 3rd postoperative day. Radiological imaging studies revealed no signs of recurrence during the 18-month postoperative follow-up.

Copyright © 2013, Buddhist Compassion Relief Tzu Chi Foundation. Published by Elsevier Taiwan LLC. All rights reserved.

1. Introduction

Mediastinal neurogenic tumors are a group of heterogeneous neoplasms derived from the tissue of the neural crest, including cells of the peripheral, autonomic, and paraganglionic nervous systems. These tumors account for approximately 20% of all mediastinal neoplasms in adults and are benign in most cases (malignancy incidence, about 3–19%) [1]. The word “paraganglioma” is applied to tumors arising from the paraganglia, regardless of their location, and they can be found in any part of the body where there are paraganglia.

Mediastinal paraganglioma (MP) originate from paravertebral and para-aortic chain ganglia [2]. Most functional MPs are noted during investigation of hypertension or other symptoms secondary to catecholamine excess. However, in the majority of cases, they are nonfunctional, and are noted incidentally, and the diagnosis is confirmed postoperatively. Many authors have reported significant bleeding during surgical removal, because MP firmly adhere to or invade adjacent mediastinal organs such as the great vessels and the heart. We report a case of nonfunctional posterior MP in a 54-year-old man that was excised successfully through a posterolateral thoracotomy.

2. Case report

A previously healthy 54-year-old man was admitted to our hospital with a 3-month history of left-side chest pain and cough. He had no other specific complaints such as palpitations, sweating, or headache. Results of physical examination and laboratory data were all within normal limits. Chest radiography showed a left side paravertebral well-circumscribed mass (Fig. 1A). Chest computed tomography revealed a left posterior mediastinal solid tumor ($6.0 \times 5.0 \times 4.0$ cm) with strong enhancement in the paraspinal region at the T7–T9 vertebral level (Fig. 1B).

Chest magnetic resonance imaging (MRI) noted no direct vertebral invasion or spinal cord compression, but the mass showed central necrosis and strong inhomogeneous gadolinium enhancement with a presumptive diagnosis of neurogenic tumor (possible meningioma or neurinoma) (Fig. 2). Urine levels of metanephrine and normetanephrine were within normal limits.

The patient underwent a left posterolateral retropleural thoracotomy through the tumor within the left paravertebral sulcus. Intraoperatively, the patient's blood pressure remained stable at 95/75 mmHg during resection of the tumor. However, the tumor showed serious invasion of the costal periosteum, and the dissection was laborious. There was significant intraoperative bleeding of about 1000 mL (not massive) because of the abundant blood supply through several feeding collaterals from the intercostal arteries. The tumor was safely and totally removed, and the postoperative course was uneventful. Hemostasis was adequate (Fig. 3). The duration of the surgery was 200 minutes.

Conflicts of interest: none.

* Corresponding author. Department of Thoracic Surgery, Mohamed V Military University Hospital, Immeuble 29, Apartment 7, Résidence Mesk Ellil, Secteur 23, Riad, Rabat 10100, Morocco. Tel.: +212 6 61818545; fax: +212 5 37711004.

E-mail address: hassankabiri@yahoo.com (E.H. Kabiri).

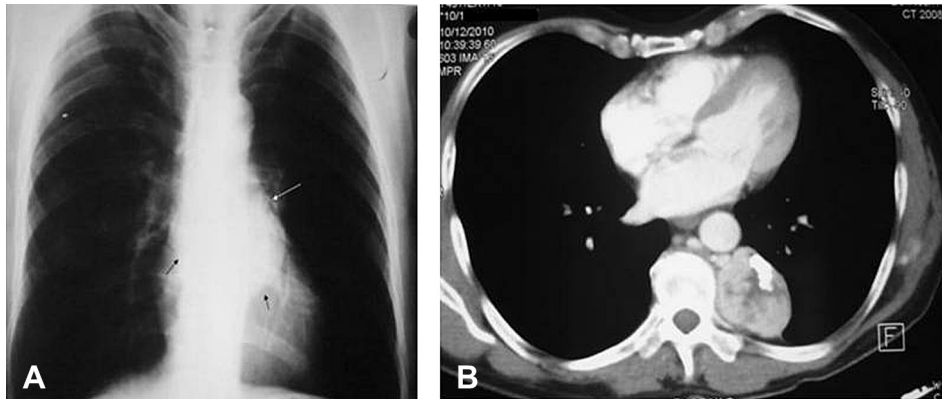


Fig. 1. (A) Chest radiograph showing a left mediastinal mass. (B) Chest computed tomography shows a large left mediastinal cystic mass within the left paraspinous sulcus (T7–T9).

The patient was discharged on the 3rd postoperative day. The pathological diagnosis showed a well-encapsulated, vascularized lesion containing proliferation of lobules separated by thin fibrous tracts, abundant eosinophilic granular cytoplasm with nests of cells, slightly hyperchromatic pleomorphic nuclei, and finely granular chromatin in an organoid pattern (HE $\times 40$) (Fig. 4). The tumor tested positive for synaptophysin, S-100 protein (Fig. 5A), and chromogranin A (Fig. 5B) but negative for cytokeratin, suggestive of a benign paraganglioma. The patient was free of recurrence after 18 months of follow-up.

3. Discussion

Paragangliomas are tumors that arise from the chromaffin cells of the sympathetic nervous system. When the tumor arises from the adrenal medulla, it refers to a pheochromocytoma; however, paragangliomas and chemodectomas are tumors arising from the chromaffin cells at a site [3]. We found fewer than 17 reports in PUBMED using the MeSH Terms (“paraganglioma”) AND (“mediastinum” OR “mediastinal”). Several were case reports, and there were very few retrospective reviews. We found no precise incidence rate of paraganglioma in the posterior mediastinum, but Ayala-Ramirez et al [4] reported 13 mediastinal locations among 104 paragangliomas.

The mediastinum is a less commonly involved site for extra-adrenal catecholamine-secreting and nonsecreting paraganglioma (2% of paragangliomas) [3]. The most commonly

encountered posterior mediastinal tumors are neurogenic tumors (75%), and the remaining 25% include a heterogeneous group of rare tumors including lymphoma, teratoma, sarcoma, and other lesions arising from other structures and involving the posterior mediastinum [5].

Functional MPs are usually diagnosed when investigating hypertension or symptoms that are a result of catecholamine secretion such as palpitations, sweating, headache, or symptoms related to compression that leads to hoarseness, dysphagia, dyspnea, and chest pain [6]. However, 17% of patients with secreting tumors remain asymptomatic, which is a diagnostic challenge [7]. The diagnosis is incidental in more than 50% of cases [8]. Nonfunctional MPs are discovered incidentally, and the diagnosis is established postoperatively by pathological study. The diagnosis of a paraganglioma is based on clinical symptoms, imaging tests, and urinary essays of catecholamine metabolites [2]. Inactive MP may present as a large tumor with local invasion or compression of adjacent structures (intercostals nerve, heart, spinal cord). The chest radiograph may show involvement of the mediastinum, a well-circumscribed mediastinal mass, or pulmonary metastasis. The characteristics of MP are typical on computed tomography scan and MRI, with a location in the bifurcation of the great vessels and intense homogeneous enhancement except for necrotic areas. Therefore, intermediate signal intensity on T1 and high signal intensity on T2 are noted. MRI can further define the degree of extension of the mass into the neural canal, and the presence of thoracic vertebral destruction or spinal cord compression [9]. The

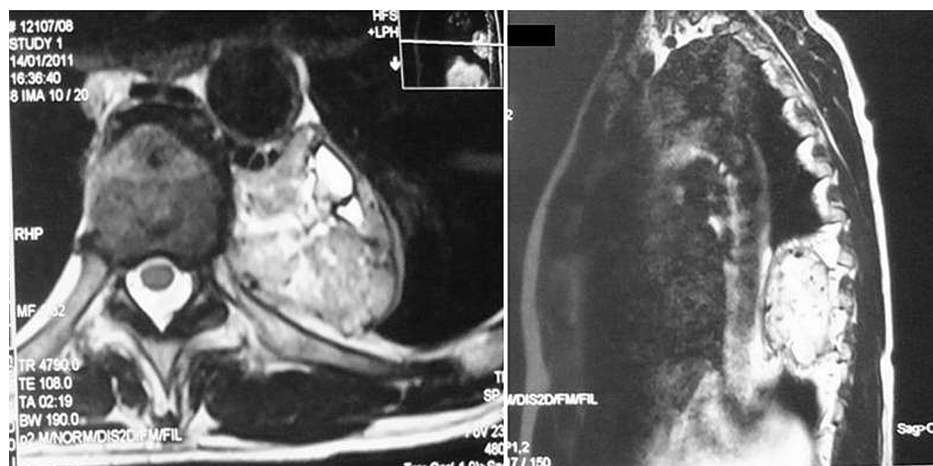


Fig. 2. Magnetic resonance imaging shows central necrosis, strong inhomogeneous gadolinium enhancement and no direct vertebral invasion or spinal cord compression.

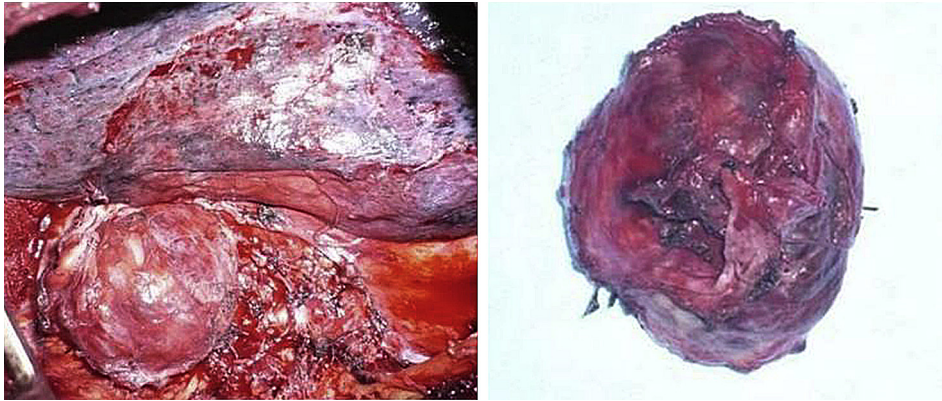


Fig. 3. Operative view of the mediastinal mass tumor.

treatment of choice for MP is surgical excision, because these tumors are resistant to chemotherapy and radiotherapy [2]. Because of intraoperative bleeding and secretion of catecholamines in patients with metabolically active tumors, surgical removal should be

complete. In the study of Ayala-Ramirez et al [4], the incidence of metastasis was higher in patients with primary paraganglioma located within the mediastinum (69%) [4].

The two major challenges in surgery of MP are intraoperative bleeding due to the hypervascular nature of the tumor and proximity to the great vessels, and systemic anticoagulation in cases of cardiopulmonary bypass. Because the tumor is highly vascularized and extensively adherent to the costal periosteum, the procedure is done through a posterolateral thoracotomy to allow satisfactory exposure, adequate bleeding control, and efficient dissection. Some authors have initially approached a well-defined mediastinal tumor using video-assisted thoracic surgery. Other authors recommend preoperative angiographic embolization to reduce the risk of bleeding, but we think that this procedure is not systematic.

Hormonal crises, although uncommon, have been associated with significant morbidity and mortality [8]. Alpha-adrenergic blocking agents are prescribed preoperatively to ensure blood pressure control and to prevent these crises [3]. Rigorous hemodynamic control and meticulous surgical technique are the key steps in the prevention and management of these complications [2].

In summary, nonfunctional MPs are very rare tumors and should be included in the differential diagnosis of mediastinal masses, particularly hypervascular tumors. Complete resection is associated with excellent survival. The patient should be carefully followed-up to detect possible recurrence or metastasis.

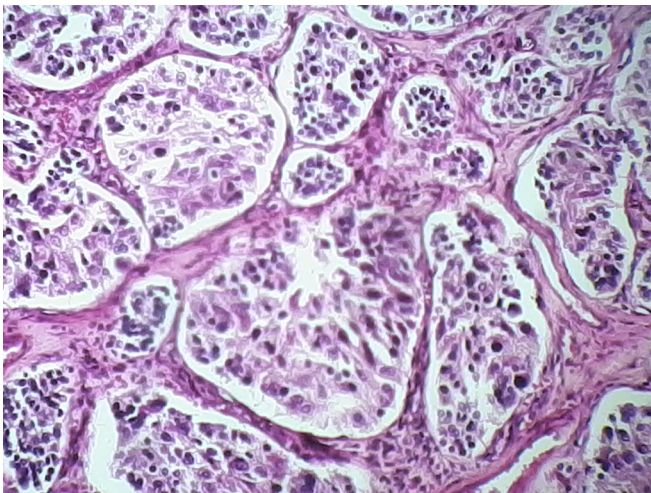


Fig. 4. Pathologic view of a proliferation of lobules separated by thin fibrous tracts (hematoxylin and eosin, $\times 40$).

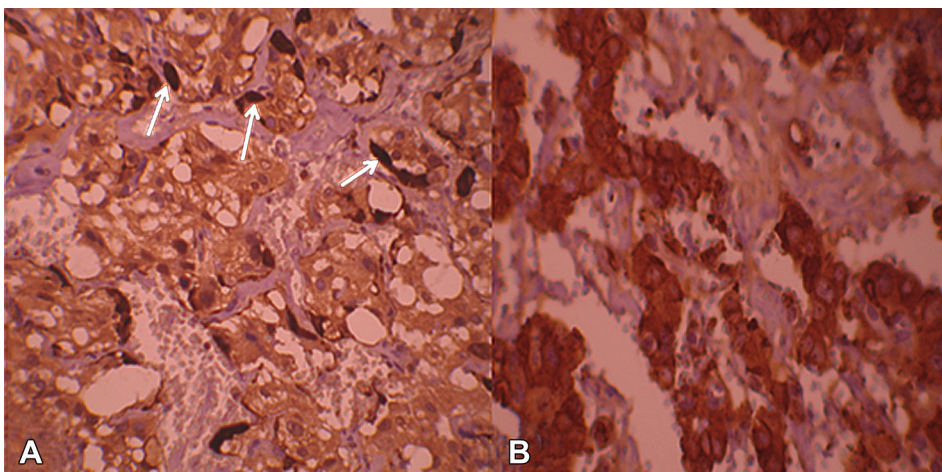


Fig. 5. Immunohistochemistry view showing intensive positivity of PS-100 in the (A) sustentacular cells and (B) chromogranin in the cytoplasm.

References

- [1] Topçu S, Alper A, Gulhan E, Koçyigit O, Tastepe I, Cetin G. Neurogenic tumors of the mediastinum: a report of 60 cases. *Can Respir J* 2000;7:261–5.
- [2] Wald O, Shapira OM, Murar A, Izhar U. Paraganglioma of the mediastinum: challenges in diagnosis and surgical management. *J Cardiothoracic Surg* 2010;5:19.
- [3] Francis IR, Korobkin M. Pheochromocytoma. *Radiol Clin North Am* 1996;34:1101–12.
- [4] Ayala-Ramirez M, Feng L, Johnson MM, Ejaz S, Habra MA, Rich T, et al. Clinical risk factors for malignancy and overall survival in patients with pheochromocytomas and sympathetic paragangliomas: primary tumor size and primary tumor location as prognostic indicators. *J Clin Endocrinol Metab* 2011;96:717–25.
- [5] Davidescu M, Motaş N, Bluoss C, Motaş C, Rus O, Horvat T. Neurogenic tumors of the posterior mediastinum. *Chirurgia (Bucur)* 2011;106:199–203.
- [6] Brown ML, Zayas GE, Abel MD, Young Jr WF, Schaff HV. Mediastinal paragangliomas: the Mayo Clinic experience. *Ann Thorac Surg* 2008;86:946–51.
- [7] Liu L, Mei J, Che G. Asymptomatic paraganglioma of the posterior mediastinum misdiagnosed until operation. *Thorac Cardiovasc Surg* 2010;58:302–4.
- [8] Young Jr WF. Paragangliomas: clinical overview. *Ann N Y Acad Sci* 2006;1073:21–9.
- [9] Balcombe J, Toigian DA, Kim W, Miller Jr WT. Cross sectional imaging of paraganglioma of the aortic body and other thoracic branchiomeric paraganglia. *AJR Am J Roentgenol* 2007;188:1054–8.