

Nonfunctional para-aortic paraganglioma: About a rare mediastinal mass

Bennani H ^{1,*}, Lkharrat F Z ¹, Bennani M M ³, Akkamar A ², Bouardi N ¹, Haloua M ², Alaoui Lamrani M Y ¹, Boubbou M ², Maâroufi M ¹ and Alami B ¹

¹ Department of Radiology, CHU Hassan II, Fès, Morocco.

² Department of Mother and child radiology, CHU hassan II, Fès, Morocco.

³ Université libre de bruxelles. Brussels, Belgium.

World Journal of Advanced Research and Reviews, 2023, 19(02), 067–071

Publication history: Received on 01 July 2023; revised on 26 August 2023; accepted on 29 August 2023

Article DOI: <https://doi.org/10.30574/wjarr.2023.19.3.1603>

Abstract

Objectives: Our purpose is to illustrate the radiological aspects of mediastinal paragangliomas.

Material: we report the case of a 73-year-old woman, who presented to the emergency department with a thoracic trauma, and to whom the diagnosis of a mediastinal mass was discovered incidentally.

Results: The patient presented to the emergency department in the context of an injury assessment of a thoracic trauma.

The patient reported a history of diabetes, but did not report any chest or cardiac symptoms.

A thoracic CT scan was performed and showed a mass in the anterior and middle mediastinum with vivid enhancement.

A mediastinoscopy with biopsy was performed and came back in favor of a paraganglioma.

This case highlights the importance of imaging in the diagnosis of mediastinal paragangliomas as a possibility to consider in the differential diagnosis of an intra-thoracic mass.

Conclusion: Paragangliomas are neuroendocrine tumors that rarely arise in the mediastinum. A good knowledge of radiological semiology coupled with scintigraphy is essential for a good diagnostic approach. However, surgery is the only effective way to diagnose and treat this pathology

Keywords: Paraganglioma; Imaging; Mediastinal mass

1. Introduction

Paragangliomas are rare neuroendocrine tumors that develop from the sympathetic or parasympathetic ganglia.

Mediastinal paragangliomas are very rare and accounts for less than 2 percent of all locations [1].

Paragangliomas can be non-functional or secreting catecholamines. Non-functional paragangliomas are the most frequent, which explains their incidental discovery at an advanced stage in up to 50 % of the cases [2], [3].

* Corresponding author: Bennani H

Imaging associated with scintigraphy plays an important role in the diagnosis and the assessment of loco-regional extension [4], [5].

Surgical resection is the treatment of choice in the management of this tumor with high survival rates with appropriate medical care [6].

2. Observation

73-year-old woman with no previous history presented to the emergency department with a thoracic trauma.

On presentation, the patient's vital signs were stable, with a blood pressure of 120/70 mm Hg, heart rate of 76, and no dyspnea.

Physical examination was normal, with no evidence of pleural effusion or parietal ecchymosis.

In the immediate aftermath, the patient underwent a spontaneous thoracic CT- scan that showed an incidental anterior mediastinal mass. **(Figure 1)**

An injection of intravenous contrast was indicated with arterial and venous phase acquisitions to characterize the lesion and define its extension.

Imaging showed an 6 cm anterior mediastinal mass located between the aorta and the superior vena cava, and adherent to the anterior tracheal wall. It presented a vivid enhancement comparable to the enhancement of the aorta in the arterial phase. **(Figure 2)**

On these semiological criterias the diagnosis of paraganglioma was suspected.

Urine vanillylmandelic acid levels were not elevated.

Therefore, the patient underwent a thoracoscopy with biopsy in favor of a paraganglioma.

Urine vanillylmandelic acid levels were not elevated.

A total body iodine-131 meta-iodo-benzylguanidine scintillation scan showed tracer accumulation in the anterior mediastinum with no sign of any further extension.

The patient was scheduled for tumor embolization followed by surgical resection.

However, the patient refused any treatment

She is still followed up in consultation without any worsening of the symptomatology.

3. Discussion

Mediastinal paragangliomas are uncommon neuroendocrine tumors that arise from sympathetic ganglia outside of the adrenal medulla [1], [2], [5].

Less than 2% of paragangliomas are located in the mediastinum [1], [5].

There are two main types of mediastinal paraganglioma; aorticopulmonary paraganglioma arising in the anterior or middle mediastinum and aortico-sympathetic paraganglioma occurring in the posterior mediastinum [1].

Paragangliomas can be non-functional or functional. Functional paragangliomas produce norepinephrine and normetanephrine which can be responsible of hypertension, and headaches [6]. The gold standard for paraganglioma diagnosis is primarily biochemical and confirmed by increased levels of 24-hour urine fractionated metanephrines and catecholamines [7].

Non-functional paragangliomas have a late onset of symptoms related to the mass effect, which is why they are often discovered incidentally at an advanced stage [2], [7]. The compression and mass effect can be manifested by chest pain, dyspnea, cough, dysphagia, hemoptysis, and superior vena cava syndrome [7], [8].

The differential diagnosis for a patient presenting with an anterior mediastinal mass is extensive and includes lymphadenopathy, thymic tumors, and teratomas.

Imaging plays an important role in the diagnostic approach to paraganglioma. In fact, On CT a paraganglioma has avid contrast enhancement comparable to the enhancement of the aorta and a delayed washout secondary to its vascularity [5]. It frequently shows heterogeneous enhancement with central latency.

Furthermore, MRI plays an important role in the characterization of the tumor as well as in understanding the loco-regional assessment and the anatomical relations with the adjacent structures allowing the resectability assessment.

It shows a tumor hypointense or isointense on T1-weighted MRI imaging and hyperintense on T2-weighted MRI imaging with variable contrast enhancement and areas of central necrosis or internal hemorrhage. [5]

31-MIBG scintigraphy is used for both localization and staging of neuroendocrine tumors, it also allows the detection of secondary localizations in malignant forms. The images must be interpreted with caution, since there are risks of false-negatives (drugs interfering with the radio-isotope) and false positives. [5]

Moreover, the functionality of paragangliomas may be elucidated through single photon emission- computed tomography [5], [9].

Surgical resection is the treatment of choice in the management of this tumor with high survival rates with appropriate medical care [6].

Patients may undergo arterial embolization to reduce the risks of operative hemorrhage. In addition, the use of perioperative alpha-blockers is essential to prevent hypertensive crisis. [6], [9]

In all patients, life-long surveillance for recurrence should be performed with regular checkups employing biochemical testing for fractionated metanephrines and catecholamines and imaging techniques. [5]

The prognosis is usually favorable after a complete resection.



Figure 1 Axial non enhanced CT scan showing an anterior mediastinal which is located close to the mediastinal structures

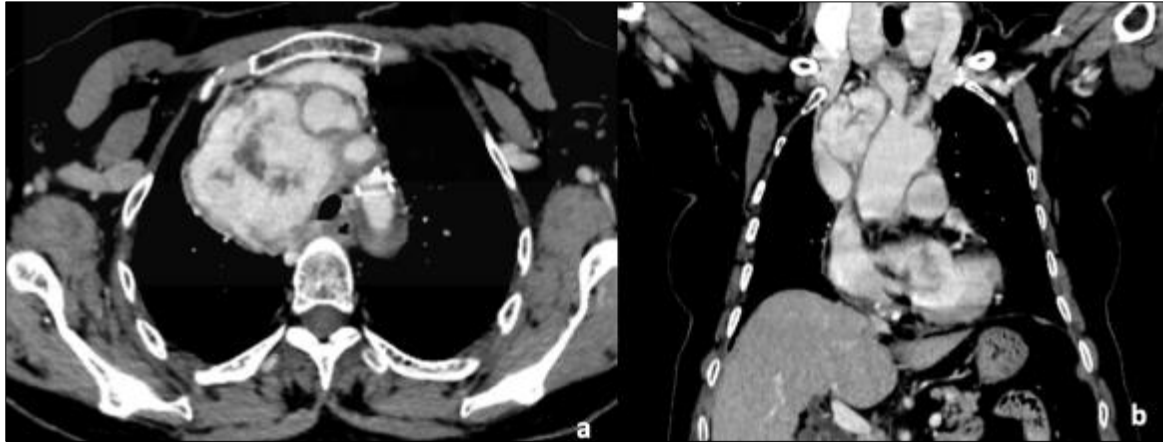


Figure 2 Axial (a) and coronal (b) enhanced CT scan (in the arterial phase) showing an anterior mediastinal mass located between the aorta and the superior vena cava, and adherent to the anterior tracheal wall, presenting a vivid on the arterial phase

4. Conclusion

Paragangliomas are neuroendocrine tumors that rarely arise in the mediastinum. A good knowledge of radiological semiology coupled with scintigraphy is essential for a good diagnostic approach. However, surgery is the only effective way to diagnose and treat this pathology

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

If studies involve use of animal/human subject, authors must give appropriate statement of ethical approval. If not applicable then mention "The present research work does not contain any studies performed on animals/humans subjects by any of the authors".

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

- [1] Y. Takashima et al., « Mediastinal paraganglioma », *Jpn J Radiol*, vol. 33, no 7, p. 433-436, juill. 2015, doi: 10.1007/s11604-015-0436-z.
- [2] M. C. B. da Silva et al., « Paraganglioma: An Uncommon Cause of Mediastinal Mass », *Circ: Cardiovascular Imaging*, vol. 13, no 2, p. e009693, févr. 2020, doi: 10.1161/CIRCIMAGING.119.009693.
- [3] O. Wald, O. M. Shapira, A. Murar, et U. Izhar, « Paraganglioma of the mediastinum: challenges in diagnosis and surgical management », *J Cardiothorac Surg*, vol. 5, no 1, p. 19, déc. 2010, doi: 10.1186/1749-8090-5-19.
- [4] F. Melillo, R. Spoladore, et A. Margonato, « Multimodality imaging of aortopulmonary paraganglioma », *European Journal of Internal Medicine*, vol. 66, p. e1-e2, août 2019, doi: 10.1016/j.ejim.2019.05.005.
- [5] N. C. Cleveland, D. N. Nguyen, K. Goel, C. D. Tran, et J. S. Mueller, « Cross-Sectional Imaging and Thoracoscopic Resection of Functional Intrathoracic Paraganglioma of the Aortic Body », *Current Problems in Diagnostic Radiology*, vol. 49, no 6, p. 489-492, nov. 2020, doi: 10.1067/j.cpradiol.2018.08.011.

- [6] M. L. Brown, G. E. Zayas, M. D. Abel, W. F. Young, et H. V. Schaff, « Mediastinal Paragangliomas: The Mayo Clinic Experience », *The Annals of Thoracic Surgery*, vol. 86, no 3, p. 946-951, sept. 2008, doi: 10.1016/j.athoracsur.2008.04.105.
- [7] C. F. Andrade, S. M. Camargo, M. Zanchet, J. C. Felicetti, et P. F. G. Cardoso, « Nonfunctioning paraganglioma of the aortopulmonary window », *The Annals of Thoracic Surgery*, vol. 75, no 6, p. 1950-1951, juin 2003, doi: 10.1016/S0003-4975(02)04841-5.
- [8] S. Szabados, E. Varady, et L. Göbölös, « Paraganglioma of the aortopulmonary window », *European Heart Journal*, vol. 30, no 10, p. 1286-1286, mai 2009, doi: 10.1093/eurheartj/ehn616.
- [9] D. M. Nguyen, J. N. Gonzalez, et N. R. Villamizar, « Surgical management of mediastinal paraganglioma: All hands on deck! », *J Card Surg*, vol. 35, no 8, p. 2047-2049, août 2020, doi: 10.1111/jocs.14746.