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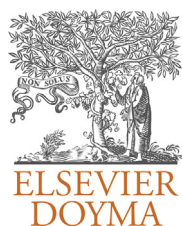
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LETTERS TO THE EDITOR

Thoracic paraganglioma diagnosed in a patient with pulmonary tuberculosis – Case report



Paraganglioma torácico diagnosticado num paciente com tuberculose pulmonar – descrição de caso clínico

Dear Editor,

Paragangliomas are rare tumors with an incidence of 0.3–0.95% of neuroendocrine tumors, which arise from

specialized neural crest cells associated with autonomic ganglia (paraganglia) 1. Nearly 90% of these tumors arise from the medulla of the adrenal gland and are called “pheochromocytomas”. When they have an extra-adrenal origin, they are termed “paragangliomas”. Intrathoracic location is infrequent, corresponding only to 10% of paraganglioma cases.¹

Herein the authors described the case of a 50-year-old male patient, Caucasian, smoker. As a background he had arterial hypertension and discoid lupus erythematosus. His usual medication was candesartan 16 mg, deflazacort 3 mg/day and chloroquine 250 mg/day. He presented in our hospital complaining of dyspnea, cough, asthenia and weight loss with three weeks evolution. His physical exam was

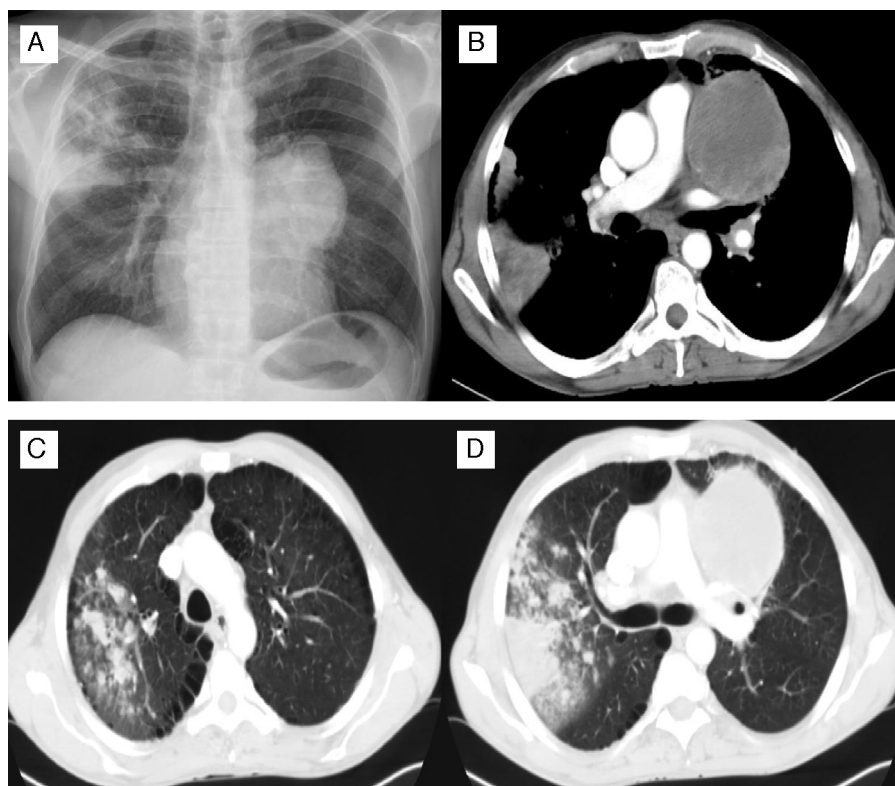


Figure 1 (A) Chest radiography: heterogeneous infiltration in the right upper lobe and homogeneous opacity in the left hilar region. (B)–(D) Chest Computed Tomography of the thorax: hypodense mass of 9 cm with well defined margins located in the mediastinum with extension to the left upper lobe, areas of consolidation in the right upper lobe associated with micro-nodular infiltration and ground-glass opacities.

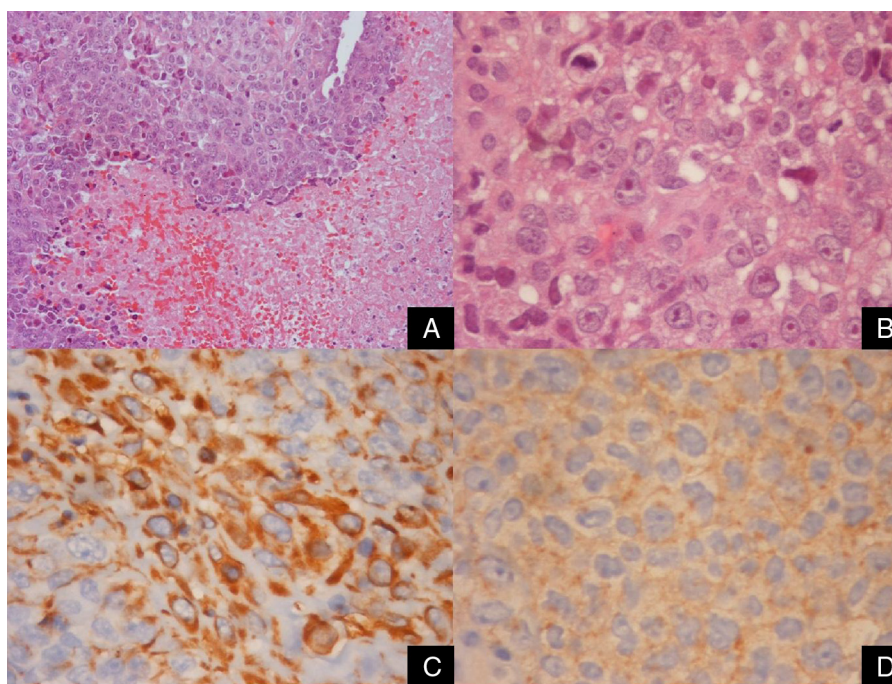


Figure 2 Surgical lung biopsy. A solid and poorly differentiated malignant neoplasm with extensive areas of necrosis (A – HE 200 \times). The tumor cells had abundant eosinophilic cytoplasm and round nuclei with evident nucleoli (B – HE 600 \times). Immunohistochemically, the neoplastic cells focally expressed vimentin (C – 600 \times) and synaptophysin (D – 600 \times) in the absence of epithelial and other markers (AE1/AE3, CK8/18, CK7, CK20, CK5, 34 β 12, EMA, TTF1, napsin-A, p63, calretinin, pS100, CD31, CD34, PLAP, beta-HCG, CD45, CD20, CD10, inhibin, C-kit, CD99, actin, desmin, GFAP, CD30, CD21, HMB45, CD56, CD23, alpha-fetoprotein, myeloperoxidase, lysozyme, WT1, ALK, HepPar-1). With those morphological and immunohistochemical characteristics in the appropriate clinical context, the diagnostic hypothesis of paraganglioma/pheochromocytoma was suggested.

unremarkable, except for malar and cervical cutaneous rash related to his discoid lupus. Chest X-ray and computed tomography showed pulmonary infiltrations in the right upper lobe and a mediastinic mass of 9 cm (Fig. 1). Abdominal CT did not show any abnormalities. Sputum analysis confirmed a diagnosis of pulmonary tuberculosis and the patient started anti-tuberculosis treatment. His HIV and hepatitis virus status were negative. Bronchoalveolar samples were negative for malignancy and CT-guided transthoracic core biopsy of the mediastinal mass only showed necrotic tissue, so the patient underwent a surgical lung biopsy.

Despite treatment, clinical evolution was not favorable, with persistent fever, one episode of auricular flutter and hemodynamic instability. Antibacillary susceptibility testing revealed resistance to all first class drugs. At the 45th day of admission, patient developed an acute pulmonary edema with no response to medical treatment and he eventually died. Description of the surgical biopsy was only available post-mortem and suggested the diagnosis of a pheochromocytoma/paraganglioma – Fig. 2.

This clinical case illustrates a patient with concomitant diagnosis of pulmonary tuberculosis and a thoracic paraganglioma. Because the results of surgical biopsy were only known post-mortem, additional research on the evaluation of the diagnosis of paraganglioma, including determination of catecholamine and metanephrine levels, presence of distant metastasis and genetic testing were not possible.

Because this is a rare tumor, most of the literature describes only single case reports.^{2,3} In 2008, Mayo Clinic reported their experience with fourteen cases of treated mediastinal paragangliomas.⁴ Concomitant occurrence of tuberculosis and thoracic paraganglioma is even rarer, with very few previous reports⁵, making the diagnosis and treatment of these cases difficult and challenging.

In conclusion, paragangliomas are not usually suspected in the initial evaluation of a mediastinal mass because of its rarity, but it should be part of the differential diagnosis. The concomitant occurrence of tuberculosis may pose a diagnostic challenge in this case, leading to a delay in the diagnosis of this type of tumors.

Ethical considerations

Protection of people and animals. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors declare that no patient data appear in this publication.

Conflict of interest

The authors declare that they have no conflicts of interest to the contents of the manuscript.

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Ethical limits for noninvasive ventilation prescription



Limites éticos para a prescrição de ventilação não invasiva

To the Editor:

Non-Invasive Ventilation (NIV) is the treatment of choice in acute respiratory failure (ARF) related to Chronic Obstructive Pulmonary Disease exacerbation and Acute Cardiogenic Pulmonary Edema.^{1,2} It has also demonstrated good results in a set of other consensual and systematized pathologies.^{1,2} In addition NIV has been used as an alternative for ARF patients who have “do-not-intubate” orders either due to poor prognosis associated with multiple comorbidities or terminal disease, or as palliative management of dyspnea.²

Aiming to encourage discussion around this theme, the authors reviewed 508 medical records of patients undergoing NIV between November 2011 and May 2013, in *Hospital Geral do Centro Hospitalar e Universitário de Coimbra*, and identified 15 cases in which the use of NIV was the subject of ethical considerations: 6 patients with advanced cancer disease, 5 with multiple organ dysfunction, 3 with extensive stroke damage, and one patient with respiratory failure (RF)

of central origin. The patients, 4 male and 11 females, had an average age of 65 ± 15 , and had an average age-adjusted Charlson comorbidity index of 6.5 ± 3.1 . Five patients presented type 1 RF, and 9 had type 2 RF. NIV was administered to one patient despite a lack of RF criteria. The average $\text{PaO}_2/\text{FiO}_2$ ratio was 187.9 ± 60.2 , and average PaCO_2 was 58.3 ± 25.0 mmHg. Seven patients presented acidosis (average pH: 7.19 ± 0.13): mixed in 4 cases, respiratory in 2, and metabolic in one patient. The application of pressure support ventilation of 11.4 ± 2.8 cmH₂O, with $50.4 \pm 21.7\%$ FiO_2 led to improved pH, $\text{PaO}_2/\text{FiO}_2$ and PaCO_2 but showed no statistical significance (Table 1). After 3.0 ± 4.2 days of NIV 13 of the 15 patients died. None of the 10 patients capable of assessing the efficacy subjectively referred relief of dyspnea.

In conclusion, we do not consider it appropriate to use NIV in situations where there is no legitimate justification. On the contrary: it is an inefficient and costly approach and often leads to misperceptions about end of life management.³ With our limited public health resources providing differentiated treatments to those who do not benefit from them could be considered ethically reprehensible because, as a consequence, treatment may not then be available for those who would benefit. NIV can on occasions contribute to a patient's comfort, when combined with other measures (like administration of morphine) in the appropriate institutions, but not in hospital emergency room.

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Table 1 Arterial Blood Gas (ABG) parameters evolution.

Parameters	Baseline	After NIV trial	P value
pH	7.31 ± 0.18	7.36 ± 0.11	0.600
PaCO_2 (mmHg)	58.3 ± 25.0	51.9 ± 12.7	0.507
$\text{PaO}_2/\text{FiO}_2$	187.9 ± 60.2	192.25 ± 67.2	0.882
NIV, non-invasive ventilation.			