Author's Reply

Respuesta de los autores

To the Editor:

As the authors of the letter stated,¹ the choice of surgical approach is still a controversial one, made worse by the small number of cases in the literature. Some authors advocate minimally invasive approaches: percutaneous drainage,² anterior mediastinotomy,³ or video-assisted thoracoscopy.⁴ Other authors⁵ advise against these, as they consider them incomplete or inadequate.

During the 11 years of our study, each surgeon chose the most economic and safest approach, and for us posterolateral thoracotomy is the incision with the best surgical field. (Although, a disadvantage is the need for patients to be lying on their side during the intervention, as this position is poorly tolerated in delicate patients and causes a restrictive pattern due to postoperative pain.) The results from Misthos et al⁵ support our decision, as they obtained a survival rate of 91% when performing a debridement via cervicotomy and thoracotomy, and a mortality of 50% when only performing a cervicotomy.

However, as the authors of the letter noted, the ultimate aim of the surgery is to debride abscesses and insert catheters for continued drainage. Therefore, the choice of approach depends in many cases on the surgeon's experience in relation to this pathology.

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However, we would like to emphasise that descending necrotising mediastinitis is a potentially fatal condition that requires a multidisciplinary and aggressive treatment to reduce mortality and the approach must provide an excellent view of the area, allowing a wide debridement and avoiding unnecessary injury to organs, vessels and nerves in that region.

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Superior Thoracic Outlet Syndrome Caused by a Subclavian Paraganglioma

Síndrome de estrecho torácico superior originado por paraganglioma subclavio

To the Editor:

Paragangliomas are rare neuroendocrine tumours. They originate in the neural crest and are usually found around vascular and nerve structures. Our case is of interest due to the extremely unusual location of the tumour and its clinical manifestations: thoracic outlet syndrome with neurological symptoms due to brachial plexus compression. Paraganglioma has never been described before with this aetiology.

We present the case of a 31 year old man who had been suffering from paresthesias and severe bouts of neuropathic and intermittent pain in his upper limb for three years. There was nothing remarkable in his medical history. There was no limb oedema or changes in colour and the pulse was present at all levels. The examinations in the upper thoracic outlet proved negative. There was a painful area on palpation in the right supraclavicular region.

A nuclear magnetic resonance (NMR) scan identified a tumour with clearly defined contours located behind and above the collarbone, immediately behind the right subclavian vein, in front of the middle scalene and posterior muscles. It measured $23 \times 36 \times 30$ mm in diameter (fig. 1a). A percutaneous biopsy led to a pathological diagnosis of paraganglioma. To complete the study and decide on the appropriate surgical procedure, a computed tomography (fig. 1b) was performed as well as an angiography of the supra-aortic trunk (at rest and in hyperextension). No vascular compromise or significant hypervascularity was apparent.

During surgery, the tumour was excised by a double supra- and infraclavicular approach. There were no immediate postoperative complications and the patient was discharged from hospital three days after surgery. He is still asymptomatic after three years.

Thoracic outlet syndrome (TOS) is a group of different diseases that affect the brachial plexus and/or the subclavian vessels as they pass through three related anatomical regions: the interscalene triangle, the costoclavicular space and the subpectoral space. Compression can occur at any of these levels due to different aetiologies which have little in common. There is a group of rare tumours that can cause TOS due to compression. Isolated cases have been described of desmoid tumour,¹ lipoma² or haemangioma of the first rib,³ among others. Our review of the literature found no cases of subclavian paraganglioma as a cause of TOS.

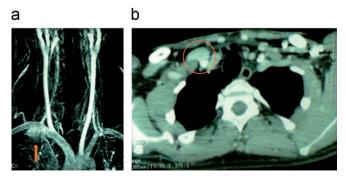


Figure 1. a) *Nuclear magnetic resonance*, which shows the intimate relationship of the tumour with the subclavian vessels. b) *Computerised tomography* showing the well-defined contours and the location of the tumour behind the collarbone.

The growth rate of paragangliomas is slow and they are usually benign. The criteria of malignancy is not defined by the histopathology of the tumour, but whether they spread to adjacent organs, due to metastasis or recurrence, which are found in 5-10% of the total.⁴ The treatment of choice is surgical resection, which is considered risky due to its anatomical location: the close relationship with the vascular-nervous structures and hypervascularity of the tumour. The main postoperative complication is sensory or motor deficit caused by nerve injury to adjacent structures. Radiation therapy is indicated for inoperable cases or as a complement to surgery after partial resection. However, this therapy does not usually completely eradicate the tumour.⁵

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Idiopathic Pulmonary Haemosiderosis in Childhood: A Good Response to Systemic Steroids, Inhaled Hydroxychloroquine and Budesonide

Hemosiderosis pulmonar idiopática en la infancia: buena respuesta al tratamiento con esteroides sistémicos, hidroxicloroquina y budesonida inhalada

To the Editor:

Idiopathic pulmonary haemosiderosis (IPH) is a rare and potentially lethal cause of diffuse alveolar haemorrhage. It is characterised by the presence of changing lung infiltrates, haemoptisis and ferropenic anaemia; with no systemic or renal associated symptoms.¹² Its clinical presentation varies from fulminant haemoptisis and acute respiratory failure to insidious clinical respiratory symptoms or refractory ferropenic anaemia.^{1,3,5}

We present the case of a 10 year old boy, with no previous respiratory symptoms, with irritative cough and progressive dyspnoea of one year of evolution accompanied by haemoptisis during the last 6 months. On exploration the most outstanding symptoms were obesity (BMI 27) and paleness of skin and mucous membranes. Additional tests showed ferropenic anaemia (haemoglobin 10.3 mg/dl, haematocrit 32%). Functional renal, liver, ions, coagulation and urine tests were all normal. Sweat test results 12 meq/l. Sputum culture was negative and Mantoux test 0 mm. On chest X-ray it was possible to see thickening of the hilum with bilateral interstitial infiltration predominantly of the lower lobes. The CAT scan can be seen in annex 1. Heart studies were normal. Negative results were obtained for immunoglobulin, complement,

