to differentiate between CD and malignancy, with cases described with SUVmax between 1.4 and 7.4 in lung involvement. Given its malignant potential, biopsy should be considered in the presence of elevated SUVs (rare in multicentric disease), or hypermetabolism in large lymph node conglomerates.[8]

Videothoracoscopy is considered a feasible technique for the diagnosis and treatment of selected unicentric cases; bleeding or firm adhesions of the mass to neighbouring structures are the main reasons for conversion to thoracotomy.[9] There is the possibility of enucleation of well-demarcated lesions, avoiding major lung resection.[2,4] Talat et al. in their systematic review (n = 404) described a surgical or debulky resection rate of 77%, more frequent in unicentric (94.3%) than multicentric (38.9%) CD, with longer overall survival in unicentric (95.3%) vs multicentric disease (61.1%) and longer disease-free interval at 3 and 5 years in unicentric disease. In unresectable lesions, residual tumour, or inoperable patients, neo or adjuvant therapies[3–5] such as adjuvant radiotherapy have been described with satisfactory long-term results and excellent local control (25−50 Gy dose) in the chest.[3,6,10] In multicentric CD, surgery is not considered a first option and is reserved for diagnostic purposes. Treatment will usually be systemic based on gluocorticoids, chemotherapy or anti-IL-6 antibodies (rituximab, tocilizumab, or siltuximab) given the elevated expression of IL-6, with a worse prognosis than unicentric disease.[3]

With our case we wish to highlight the similarity of unicentric CD with lung cancer, emphasising the importance of establishing a differential diagnosis of this disease with other entities. Preoperative diagnosis is usually complex due to the nonspecific clinical and radiological manifestations. Surgical resection of the lesion and postoperative histopathological study is required to reach a diagnosis of certainty.

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Leiomyosarcoma of the azygos vein. An unusual case☆

Leiomiosarcoma de vena ácigos. Un caso inusal

Soft tissue sarcomas (STS) are a heterogeneous and rare group of malignant tumours of mesenchymal origin. They account for 1% of all malignant neoplasms[1] and the most common of these is leiomyosarcoma[2]. This lesion is made up of smooth muscle differentiation cells[1] and accounts for 20% of all sarcomas[2].

We present the case of a 63-year-old woman who was followed up for dorsalgia and underwent MRI of the dorsal

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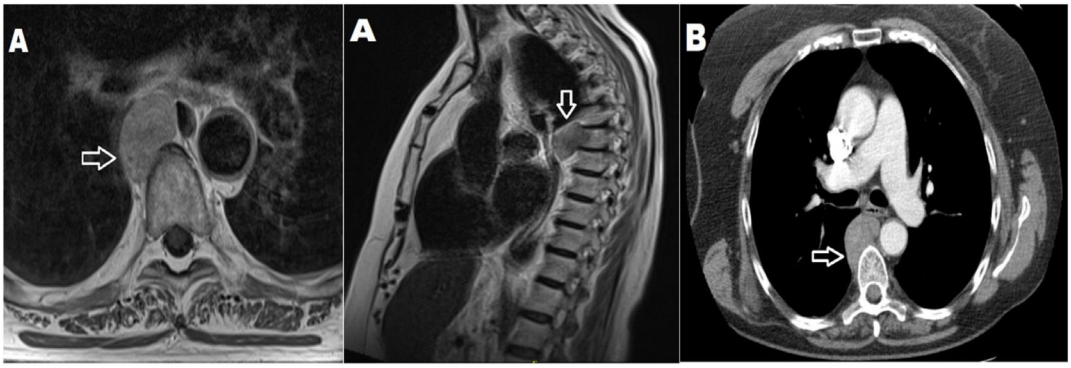

Fig. 1 – Radiologic tumour findings:
A) Contrast-enhanced MRI of the dorsal spine, axial and sagittal view: well-defined solid tumour (white arrow) in the right posterior paravertebral mediastinum at T7-T8 level, unrelated to foramen conjunctiva, causing slight remodelling of the anterior vertebral margin, without bone invasion.
B) Axial computed tomography image, mediastinal window: oval solid tumour in posterior mediastinum (white arrow).

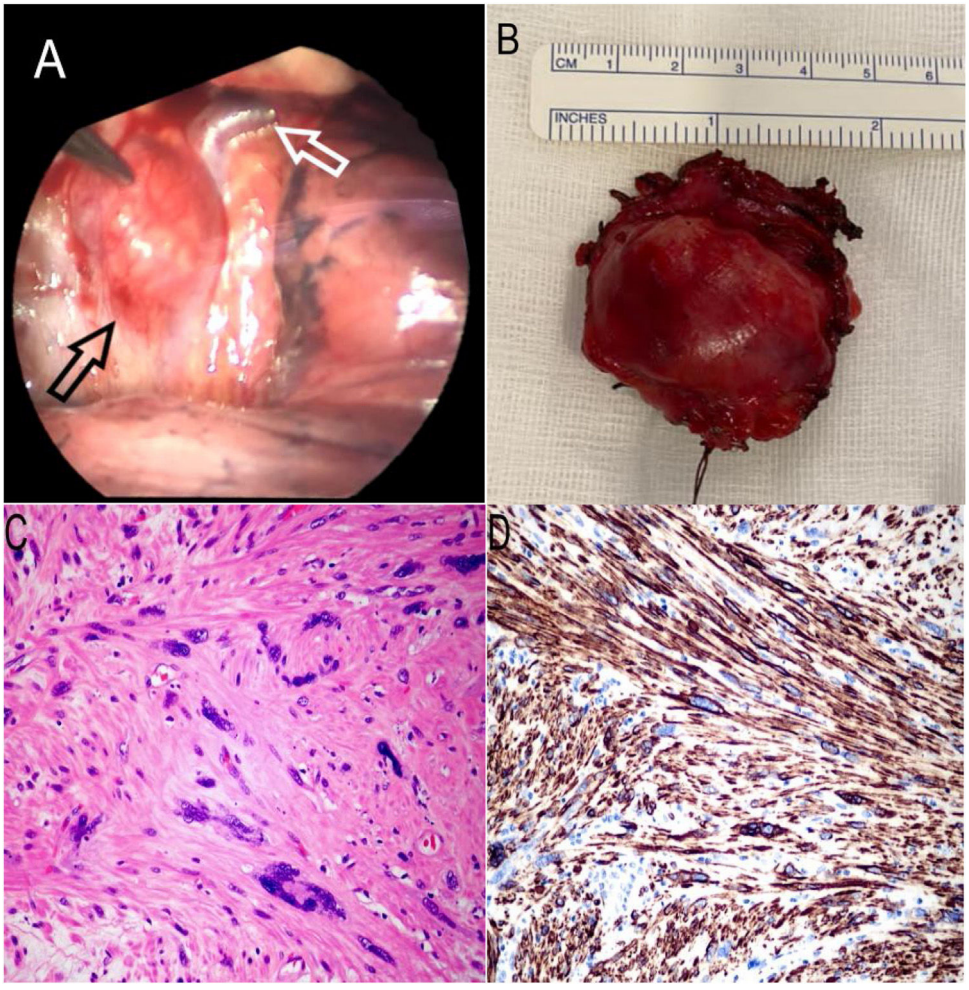

Fig. 2 – Macroscopic view and anatomopathological study.
A) Intraoperative view: hard tumour (black arrow), encapsulated, highly vascularised with nutrition from the azygos vein (white arrow).
B) Solid tumour measuring 4 × 3 cm, with an elastic consistency.
C) Microscopic study shows a malignant mesenchymal proliferation of spindle cells with fibrillary cytoplasm, blunt edges, long-linear nuclei, and others with a bizarre appearance (haematoxylin-eosin, 20×).
D) A tumour immunophenotype of muscular origin is observed, with diffuse expression of the immunohistochemical marker Desmin (20×).

spine showing a solid tumour measuring 4.4 × 4.0 × 2.4 cm. Circumscribed in the right paravertebral posterior mediastinum at the level of T7-T8 with no relation to the foramen conjunctiva (Fig. 1A). To complete the study, digestive endoscopy was performed, which excluded mucosal alteration of the oesophagus at this level, and a CT-guided biopsy with a coarse needle biopsy, which diagnosed the tumour as a low-grade leiomyosarcoma. Its origin was the azygos vein and its location was assessed on CT (Fig. 1B), which was subsequently confirmed by tumour resection.

Surgery was performed by right thoracotomy through the 5th intercostal space with preservation of the serratus anterior muscle.

The intraoperative view showed a hard, encapsulated, highly vascularised tumour with nutrition from the azygos vein and intercostal arteries, extending from the lower edge of the azygos vein arch, continuing longitudinally adjacent to the oesophagus and vertebral bodies as well as the junction of the transverse processes of the costal arches 5,6 and 7 (Fig. 2A).

To excise the tumour, dissection of the parietal and mediastinal pleura limiting the tumour was carried out, identifying the longitudinal edge with respect to the oesophagus, which respected it, as well as the vertebral bodies and transverse processes, intimately adherent but without macroscopic tumour infiltration. Dissection of the azygos vein in contact with the tumour was completed, requiring sectioning and mechanical vascular suturing, finally achieving en bloc tumour excision with the affected vein (Fig. 2B). Finally, the edges of the surgical site were marked with clips. No pathological lymphadenopathies were visualised.

The anatomopathological study confirmed the origin of the azygos vein as a grade 2 leiomyosarcoma (according to FNCLCC) with focal involvement of the soft tissue resection margin.

To complete the treatment, the clinical case was discussed at the Multidisciplinary Committee for Thoracic Tumours and the patient was referred to Radiation Oncology, receiving a total of 33 sessions on the surgical bed, with good tolerance. After this, the patient continued clinical radiological follow-up, with no evidence of tumour recurrence to date.

Leiomyosarcomas can originate in any vessel, being an extremely rare origin, accounting for <2% of all leiomyosarcomas, affecting up to 5 times more arteries than veins[3].

The most common venous origin is the inferior vena cava, which accounts for 50% of all venous leiomyosarcomas, with a total of 450 described in the literature[4].

Our case is frankly unusual due to the very low frequency of origin in the azygos vein, as to date only 2 cases have been described in the literature[5,6]. The first case was described in 1995 by Levett[5], it was a leiomyosarcoma with origin in the superior vena cava and azygos vein, and after resection it was necessary to replace it with a superior vena cava graft. The second case was described by Dasika in 1998[6], reporting a leiomyosarcoma of the azygos vein that was excised by previously ligating the azygos vein both proximally and distally, without the need for reconstruction.

Given the rarity of this tumour, the best results are obtained with a multidisciplinary approach with teams experienced in the management of these tumours.

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