Primary adrenal leiomyosarcoma: A case report☆

Leiomiosarcoma suprarrenal primario: a propósito de un caso

We present a clinical case of primary adrenal leiomyosarcoma, a soft tissue cancer originating in the muscle layer of the veins in the adrenal gland. This is an extremely rare condition with very few cases described in the literature.

The patient was a 77-year-old man whose most notable previous medical history was atrial flutter (treated by ablation) and hypertension (treated with losartan and hydrochlorothiazide, but without achieving the targeted blood pressure). Abdominal magnetic resonance imaging as part of urological investigations for prostatic hypertrophy revealed a retroperitoneal mass in the left adrenal region of 102×70 mm, considered to be a possible adrenal mass or enlarged conglomerate of lymph nodes. A subsequent chest/abdominal CT scan showed a large retroperitoneal mass with a solid appearance, measuring $70 \times 80 \times 80$ mm, which seemed to be attached to the left adrenal gland. In view of that finding, the patient was referred to the endocrinology clinic for functional tests.

There was no clear evidence in the patient's medical history or physical examination to suggest adrenal hormone dysfunction. There was also no evidence of lower back pain or of abnormal venous return. The adrenal function studies were as follows: late-night salivary cortisol <0.054 μg/dl (normal: <0.208 μg/dl), post-dexamethasone overnight salivary cortisol 2.1 μg/dl (normal: <1.8 μg/dl), renin 42.1 μU/mL (normal: 2.8–39.9 μU/mL), aldosterone 5.54 ng/dl (normal: 1.17–23.6 ng/dl), metanephrine 73 pg/mL (normal: <100 pg/mL), normetanephrine 145 pg/mL (normal: <180 pg/mL). These results suggested the diagnosis of a non-functioning adrenal mass.

As it was a large adrenal mass, it was decided to operate and considered unnecessary to present the case to the multidisciplinary committee as per treatment guidelines. No other diagnostic tests were performed prior to the intervention.

En bloc resection was performed of the left retroperitoneal mass with the left kidney, as the renal pedicle was surrounded by the mass and could not be extricated. The mass was defined in the surgical report as solid, nodular, hard and fixed, with the appearance of sarcoma.

The pathology study (Fig. 1) identified a 101×82 mm tumour attached to the left adrenal gland with free margins in the macroscopic analysis. Histological analysis showed it to be a tumour arising from the muscularis of the vessel, moderately differentiated and without necrosis. It was reported as a well-defined cell proliferation made up of spindle-cell elements with atypia in a storiform pattern, observing a maximum of seven mitoses per 10 high-power fields. Ki67 was 5%–10%. A broad IHC panel was performed with the following results: negative for oestrogen receptors, synaptophysin, chromogranin, alpha-inhibin, CD34, S100, D240, EMA, broad-spectrum cytokeratin, calretinin and STATE6; positive for smooth muscle actin and caldesmon. Surgical borders not affected. Sarcoma score: differentiation (2), necrosis (0) and mitosis (1) = total score 3 (grade 1). Pre-aortic lymph nodes showed no evidence of malignancy. Final diagnosis of leiomyosarcoma with infiltration.

In the subsequent follow-up, an oncological 18F-FDG PET/CT was performed, reporting postoperative changes in the abdomen, with no evidence of hypermetabolic foci suggestive of malignancy. Three months after surgery the patient is asymptomatic and the absence of adrenal insufficiency has been confirmed.

Adrenal incidentaloma is a common reason for consultation in endocrinology, with a growing incidence due to the greater use of imaging techniques. In radiological tests, a rate of 3% is estimated in 50-year-old patients, which can reach up to 10% in older patients.[1]

Leiomyosarcomas are a subtype of soft tissue sarcoma that develop in the smooth muscle contained in various organs such as the uterus or the digestive tract. However, there are very few recorded cases of primary adrenal leiomyosarcoma. It is believed to originate from the smooth muscle wall of the central vein of the adrenal gland and its branches.[2]

In the few reported cases of primary adrenal leiomyosarcoma, no differences have been observed in the incidence between genders or in the laterality of the tumour, which is unilateral in most cases. The age range is very broad, with a higher prevalence among people in their fifties. It has been suggested that it may be related to immunosuppression, and to positive serology for HIV and Epstein-Barr virus.[3]

The symptoms are usually associated with the compression of neighbouring structures due to growth of the tumour in the retroperitoneum, with ipsilateral lower back pain the most common symptom. The average size of the mass is 10 ± 5.9 cm. Vascular invasion is found in 26.5% of reported cases, and may cause lower limb oedema, angioedema or abdominal varices.[4]

Preoperative diagnosis is very difficult as there are no analytical or radiological data or other biomarkers to differentiate primary adrenal leiomyosarcoma from other possible abnormalities detected by imaging techniques. Furthermore, biopsy is not recommended unless there is a history of malignancy of another origin.[1] Leiomyosarcomas do not cause adrenal hormone hyperfunction. In view of all the above, diagnosis is obtained mainly after surgery, after histological analysis.[3,4] It is worthwhile assessing the characteristics and size of the structure described in the imaging technique, as this can modify the therapeutic approach.[1]

Pathology analysis is essential for diagnosis. The presence of spindle cells is usually described, with proliferation of crossed bundles, nuclear pleomorphism and small prominent nucleoli with moderate polymorphism, necrosis and haemorrhage.[5] Conventional leiomyosarcomas show reactivity for various soft tissue markers, such as desmin, smooth muscle actin and vimentin.[4] Although most are of the con-

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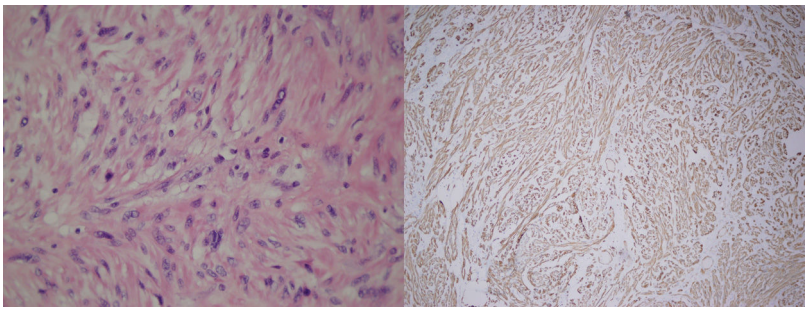

Figure 1 Left: presence of atypical spindle cell proliferation (HE 40X). Right: diffuse positivity for smooth muscle actin on IHC.

ventional type, there are pleomorphic variants, in which the expression of markers is variable.[6,7]

The treatment of choice is complete resection of the lesion. The use of adjuvant radiotherapy can be worthwhile in patients with a poor prognosis.[8]

The prognosis depends both on the size of the lesion and its extension and location, the presence of venous thrombosis, invasion of neighbouring structures and the presence of metastases, as well as the histological characteristics. Obtaining disease-free surgical margins is the greatest prognostic factor for survival. Despite having a slow growth and metastatic spread not being common until the final stages of the disease, prognosis is poor because of local recurrences.[9,10]

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