CLINICAL IMAGE

A large retroperitoneal tumor mimicking adrenocortical cancer arising in an adrenal rest

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We report a case of a 39-year-old man who complained of an observed increase in waist circumference as well as abdominal discomfort and early satiety following food intake. On physical examination, there was a palpable mass in the left epigastric region. A computed tomography (CT) scan revealed the presence of a very large retroperitoneal tumor (FIGURE 1A-1D), with no evidence of infiltration of the adjacent tissues or distant metastases. In the native phase, the neoplasm was homogenous, with one small punctate calcification. The mass was well circumscribed, without any fat stranding. Both adrenal glands were normal in size, morphology, and localization. There were no clinical or biochemical indices of abnormal hormonal activity prior to surgery. The patient underwent complete surgical resection of the tumor via conventional retroperitoneal approach.

The initial histopathologic diagnosis of the tumor (measuring 15 × 9 × 6 cm) was adrenal adenocarcinoma. The immunohistochemical profile of the lesion was as follows: melan (+), inhibin (+), cytokeratin AE1/AE2 (+/–), synaptophysin (+), chromogranin (–), and the Ki67 index <1% (FIGURE 1E and 1F). However, the absence of hormonal activity or any nearby tissue infiltration or distant metastases despite the extreme size of the tumor, the lack of a connection between the tumor and the adrenal gland on CT scans confirmed by 2 independent radiologists, extra-adrenal anatomical location found on laparotomy, the presence of normal adrenal glands on a postsurgical CT scan, and full functional hormonal reserve of the adrenal glands following surgery suggested that the tumor most probably derived from heterotopic accessory adrenal tissue present in the retroperitoneal space (adrenal rest). The tumor fulfilled only the following Weiss criteria: focal invasion of the capsule, angioinvasion of a few small vessels of the capsule, and extensive necrosis (mostly ischemic).[1] The mitotic rate was only 0–1/10 high power fields. The final consultation of histopathologic specimens resulted in a diagnosis of oncocytic adrenocortical neoplasm

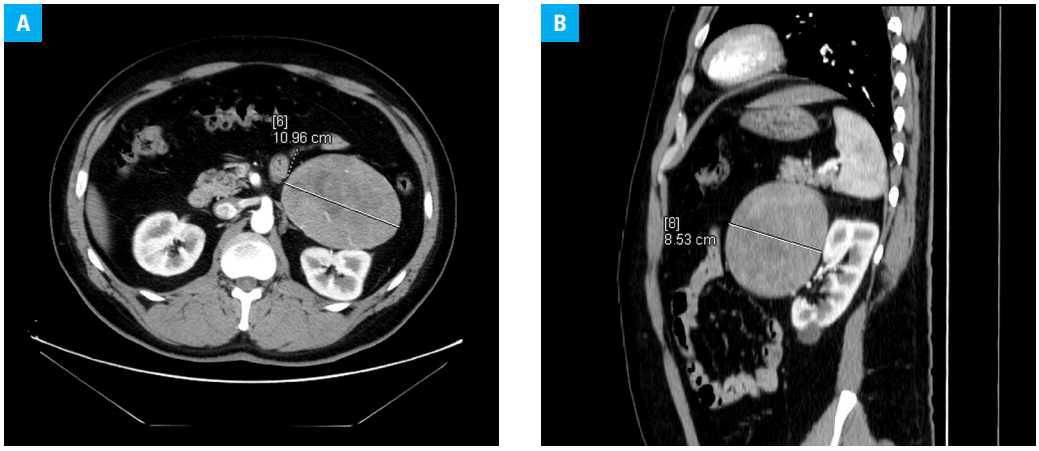

FIGURE 1 A, B – computed tomography; a large pathological soft tissue mass measuring 10.96×8.53×10.06 cm in the left retroperitoneal space

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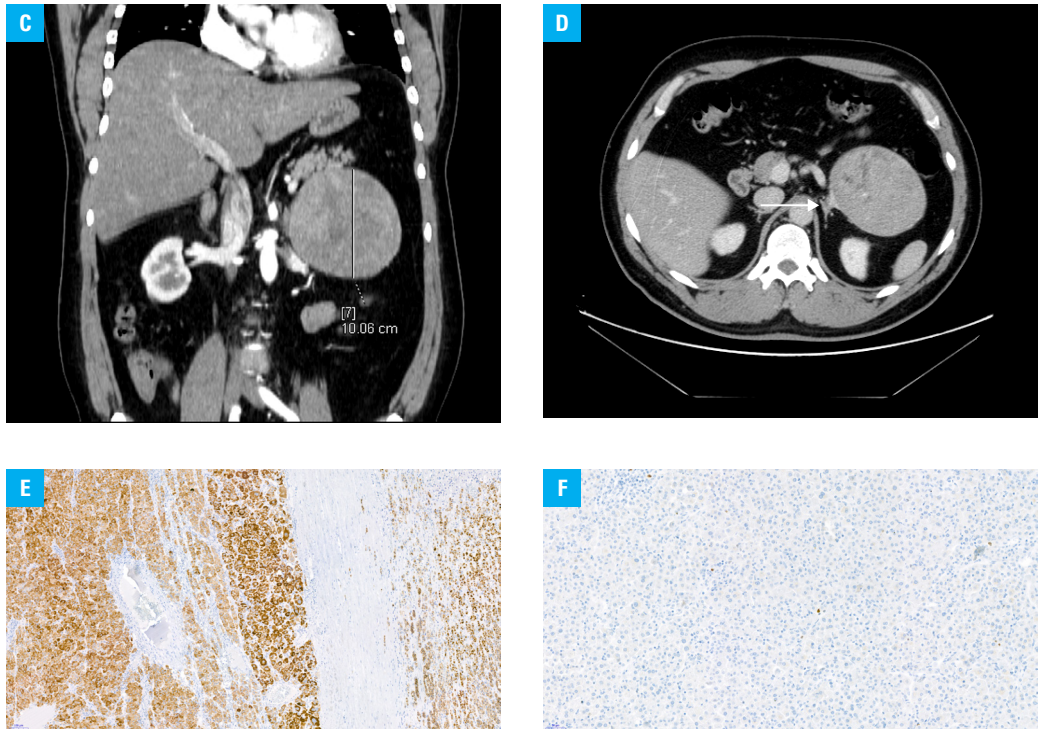

FIGURE 1 **C** – computed tomography scan showing a large pathological soft tissue mass (measuring 10.96×8.53×10.06 cm) in the left retroperitoneal space; **D** – the tumor adheres to the body and lateral limb of the left adrenal gland (arrow) (however, the adrenal gland is not enlarged), displaces the left kidney posteriorly, and elevates the tail of the pancreas. After intravenous administration of contrast media, the tumor shows slightly inhomogeneous enhancement: from 37 Hounsfield units (HUs) in the native phase to 58 HUs in the arterial phase. Enhancement is prolonged and persists in the venous phase (73 HUs). Finally, in the delayed phase, washout of contrast media is observed (63 HUs). **E** – positive immunohistochemical staining of the tumor for melan A (original magnification × 100), confirming adrenal tissue origin of the lesion; **F** – immunohistochemical staining for Ki67 index (original magnification × 200) presenting extremely low proliferative activity (<1%)

of uncertain malignant potential. The patient did not receive mitotane therapy. No recurrence or metastasis was observed during the 4 years of close monitoring and follow-up.

A wide spectrum of benign and primary or metastatic malignant tumors can be found in the peritoneum and may constitute a diagnostic challenge.[2] Although ectopic adrenal tissue is claimed to occur in 50% of neonates and children, it persists only in about 1% of adults.[3] Though tumors arising from adrenal rest tend to be asymptomatic, they may occasionally present with hormonal activity or undergo neoplastic transformation.[4] Considering a wide range of lesions that may arise in the retroperitoneal space, as well as their morphologic variability and histologic similarity, the differentiation between adrenocortical carcinoma and other benign or malignant adrenal neoplasms might be difficult.[5] The size of the tumor and its clinical presentation in terms of symptoms associated with local growth and hormonal activity might be indicative of cancer.[1,5] Additionally, adrenal cancers are in most cases functional, causing endocrinopathy.[4] The accuracy of final diagnosis is crucial for proper postsurgical management planning, including the frequency of control imaging studies, selection of adjuvant therapy (ie, mitotane treatment), as well as prognosis assessment.

ARTICLE INFORMATION

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CONFLICT OF INTEREST None declared.

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