Large retroperitoneal tumor mimicking adrenocortical cancer arising in an adrenal rest

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Article type: Clinical image

Received: February 17, 2019.

Accepted: April 10, 2019.

Published online: April 10, 2019.

ISSN: 1897-9483
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Running title: Retroperitoneal tumor

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Conflict of interest: none declared

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Here we report a 39-year-old male, who was diagnosed due to 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 1), with no evidence of infiltration of the adjacent tissues or distant metastases. In native phase neoplasm was homogenous, with one small punctuate calcification. Pathological mass was well circumscribed, without any fat stranding. Both adrenal glands were normal in size, morphology, and localization. There were no clinical nor biochemical indices of abnormal hormonal activity prior to surgery. The patient underwent complete surgical resection of the tumor via conventional retroperitoneal approach.

Initial histopathological diagnosis of the tumor sized 15x9x6 cm was adrenal adenocarcinoma. Immunohistochemical profile of the lesion was as follows: melan (+), inhibin (+), cytokeratin AE1/AE2 (+/-), synaptophysin (+), chromogranin (-), Ki67 index <1%. However, lack of hormonal activity nor any nearby tissue infiltration or distant metastases, despite the extreme size of the tumor, as well as lack of connection of the tumor and adrenal gland on CT scan pictures confirmed by two independent radiologists, extra-adrenal anatomical location found on laparotomy, presence of normal adrenal glands on postsurgical CT scan and full functional hormonal reserve of adrenal glands following surgery suggested that tumor most probably derived from accessory adrenal tissue present in heterotopic localization in the retroperitoneal space (adrenal rest). According to the Weiss criteria [1], the tumor fulfilled only the following criteria: focal invasion of the capsule, angioinvasion of a few small vessels of the capsule, extensive necrosis (mostly ischemic). Mitotic rate was only 0-1/10 high power fields. Final consultation of histopathological specimens resulted in a diagnosis of oncocytic
adrenocortical neoplasm of uncertain malignant potential. The patient did not receive mitotane therapy. There was no demonstration of recurrence or metastasis during four 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 present 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, occasionally may present with hormonal activity or undergo neoplastic transformation [4]. Taking into consideration a wide range of lesions that may arise in the retroperitoneal space, its morphologic variability and histologic similarity, the differentiation between adrenocortical carcinoma and other benign or malignant adrenal derivatives 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 [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 i.e. the frequency of control imaging studies, driving the choice of adjuvant therapy (i.e. mitotane treatment), as well as ascertaining a prognosis.
Figure 1. Computed Tomography. In left retroperitoneal space big pathological soft-tissue mass measuring 10.96x8.53x10.06 cm (width x anterior-posterior x height), (a, b and c). Tumor adheres to body and lateral limb of left adrenal gland (however adrenal is not enlarged, d), displaces left kidney posteriorly and elevates tail of pancreas. After intravenous administration of contrast media tumor shows slightly inhomogeneous enhancement - from 37 Hounsfield Units (HU) in native phase to 58 HU in arterial phase. Enhancement is prolonged and persist in venous phase (73 HU). Finally, in delayed phase washout of contrast media is observed (63 HU). Picture demonstrating positive immunohistochemical staining of the tumor for Melan A (original magnification 100x) confirming adrenal tissue origin of the lesion is presented (e). Immunohistochemical staining for Ki-67 (original magnification 200x) presenting extremely low proliferative activity (<1%).

Acknowledgements
Authors wish to acknowledge Prof. Dariusz Lange, Prof. Jan Bręborowicz, Prof. Przemysław Majewski and dr Małgorzata Janicka-Jedynska for their expert review of the specimens and discussion on the final histopathological diagnosis.

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