

Angiosarcoma of the adrenal gland

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A 55-year-old man was admitted to the Department of Internal Medicine and Metabolic Diseases, Medical University of Silesia, Katowice, Poland, in July 2012 to investigate the hormonal function of a tumor in the left adrenal gland. Two weeks prior to admission, the patient was examined by his general practitioner because of severe pain in the left lumbar region. Ultrasonography showed a nonspecific tumor located around the upper pole of the left kidney. Abdominal computed tomography performed in a municipal hospital suggested pheochromocytoma ($14 \times 14 \times 12$ cm) of the left adrenal gland (FIGURE 1ABC). On admission, the patient complained of significant weight loss in the past few weeks (a decrease of 7 cm in the waist circumference). On a physical examination, there was tenderness in the left epigastrium and mesogastrium with a palpable pathological mass. The left lumbar region was painful while shaking. Routine laboratory tests revealed anemia (9.03 g/dl), thrombocytopenia ($78.0 \times 10^9/l$), leukocytosis ($10.8 \times 10^9/l$; predominance of neutrophils in peripheral blood smear was confirmed), iron deficiency (20 $\mu\text{g}/\text{dl}$), and increased serum C-reactive protein (68.0 mg/l). An abdominal ultrasound showed an inhomogeneous area adjacent to the upper pole of the left kidney. The area measured 143×102 mm and was characterized by single calcifications and coding color flow in pulsed-wave Doppler ultrasound. Hormonal tests did not reveal hormonal activity of the tumor. The cortisol circadian profile as well as serum concentrations of androstenedione, testosterone, dehydroepiandrosterone sulfate, and the amount of metanephrine/normetanephrine in 24-hour urine sample were normal. The concentration of cortisol in the overnight 1-mg dexamethasone suppression test was slightly increased (5.75 $\mu\text{g}/\text{dl}$). The patient was urgently referred for surgical treatment and histopathology of the tissues revealed angiosarcoma.

Angiosarcomas are an extremely rare group of malignant tumors. They account for about 1% of soft tissue sarcomas, which represent 1% of all solid tumors in adults (the incidence of angiosarcoma is 0.1–0.2/100 000 per year).¹ A clinical

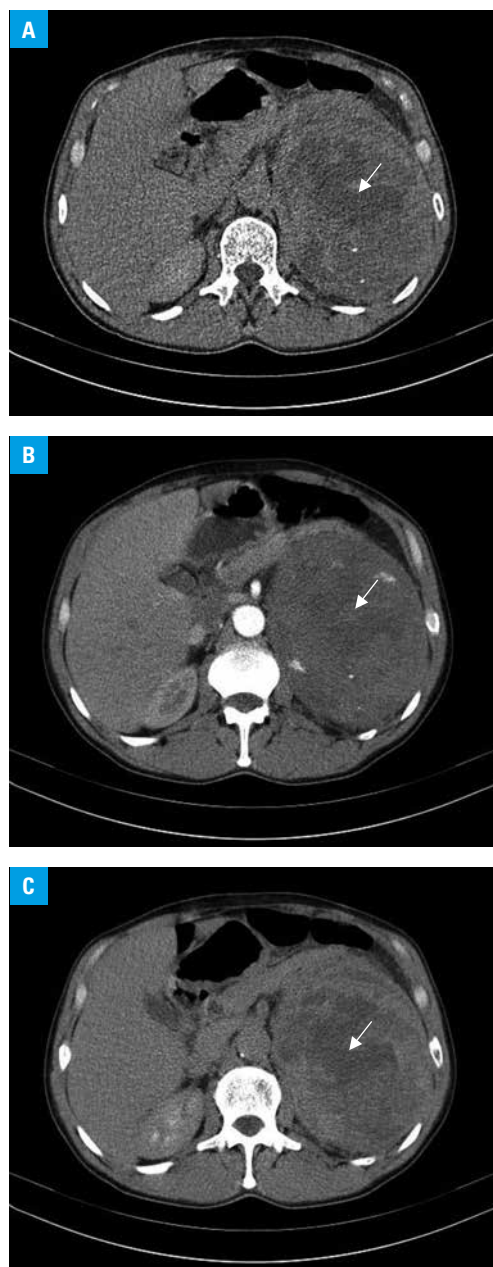


FIGURE 1 Computed tomographic images of the tumor

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picture of angiosarcomas is highly heterogeneous. They derive from the vascular endothelium and can be located at all sites in the body. Surgery, combined with chemotherapy and radiotherapy, is the treatment of choice in advanced cases. The prognosis is poor: a 5-year survival is reported in 25% to 45% of the patients.²

So far, only several cases of adrenal angiosarcomas have been reported.³ A patient with adrenal angiosarcoma can either be asymptomatic or may suffer from slight fever, anorexia, fatigue, weakness, and chronic pain in the upper abdominal quadrants. The radiological features of angiosarcoma are also indistinct so the diagnosis is supported by histopathology and immunohistochemistry.⁴ In the differentiation of tumors within the adrenal glands, we first take into consideration benign lesions (adenomas, cysts, myelolipomas, ganglioneuromas, angiomas, and hematomas), followed by malignancies (cancer, ganglioneuroblastoma), metastases (predominately from the kidney, lung, stomach, large intestine, and mammary gland), and pheochromocytoma. Yip et al.⁵ analyzed the data for 196 adrenalectomies; the presence of angiosarcoma was confirmed histopathologically only in 1 case.⁵

Our case report is the only description of such a large angiosarcoma in the adrenal glands.

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