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Leiomyosarcoma in adrenal gland

***Przemysław Dymek*[1], *Kajetan Kiełbowski*[1], *Sandra Sienkiewicz*[1], *Tomasz Błaszkowski*[1], *Aleksandra Kudrymska*[2], *Mirosław Halczak*[1], *Jerzy Lubikowski*[1], *Józef Kładny*[1]**

[1]*Department of General and Oncological Surgery, Pomeranian Medical University, Szczecin, Poland*
[2]*Department of Pathology, Pomeranian Medical University, Szczecin, Poland*

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Leiomyosarcomas are malignant neoplasms that present smooth muscle cell differentiation, which are frequently located in the retroperitoneum (12–69%) [1]. However, they may also originate from various tissues that contain smooth muscle cells. One of the least expected location of such tumours is the adrenal gland. So far, less than 50 cases have been published in the English literature. Neoplastic transformation of smooth muscle cells of the central adrenal vein or its tributaries is a prelude to the development of the tumour, often reaching large measurements. [2]

A 66-year-old male was admitted to the emergency department due to severe pain of upper right abdomen. The patient was burdened with arterial hypertension and diabetes type II. In addition, he underwent atrial cryoablation because of atrial fibrillation. Abdominal ultrasonography examination revealed a large mass located in the right adrenal gland (Fig. 1). Adrenal adenoma was the primary consideration. Therefore, an adrenal computer tomography protocol was administered. The obtained radiological imaging confirmed the presence of a tumourous mass with heterogeneous circumferential enhancement contrast. The lesion was adjacent to the hepatic lobe and upper pole of the right kidney. Abnormalities of the surrounded organs were excluded. The adrenal location of the tumour accompanied by arterial hypertension suggested pheochromocytoma. Thus, further evaluation was performed to verify this consideration. 24-hour rhythm of ACTH, cortisol, and methyl catecholamine examinations did not show any impairments. Furthermore, correct surpassing with 1 mg dexamethasone was observed. The obtained results remained unclear, and pheochromocytoma was still considered in the diagnostic process. As a result, a needle coarse biopsy could not be performed. The patient was admitted to the General and Oncological Surgery Clinic in October 2020. However, primary surgery was postponed due to atrial fibrillation. Right laparoscopic adrenalectomy was performed 2 days later. Surgery and postoperative course were uneventful, and the patient was discharged from hospital in good physical condition. Histopathological examination revealed an encapsulated (11 × 8 × 6.5 cm) mesenchymal tumour composed of spindle neoplastic cells (Fig. 2) with cytologic pleomorphism, showing immunohistochemical expression of the markers of muscle differentiation: actin, desmin, and caldesmon (Fig. 3). The cells were negative for staining of SOX10, EMA, MDM2, S100, CD10, and CD34. The proliferation index was approximately 10–15%, and the mitotic index was 3 per 10 high-power fields (HPFs). Finally, leiomyosarcoma G1 in the right adrenal gland was diagnosed. Further adjuvant therapy was not suggested by the multidisciplinary team. Follow-up computer tomography was performed 3 months postoperatively and did not reveal any recurrence.

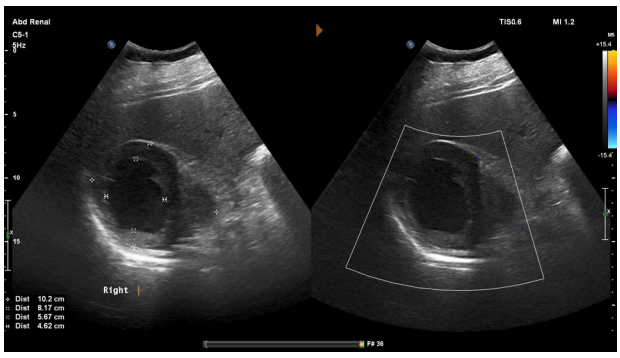

Figure 1. *Ultrasonography imaging. Heterogeneous lesion in the right adrenal area with central hypogenic area. Measurements: 10.2 × 8.17 × 5.67 × 4.62 cm*

Przemysław Dymek, Department of General and Oncological Surgery, Pomeranian Medical University, Szczecin, Poland, ul. Unii Lubelskiej 1, 71–252 Szczecin, Poland, tel: (+48) 888 477 823; e-mail: przemyslaw.dymek97@gmail.com

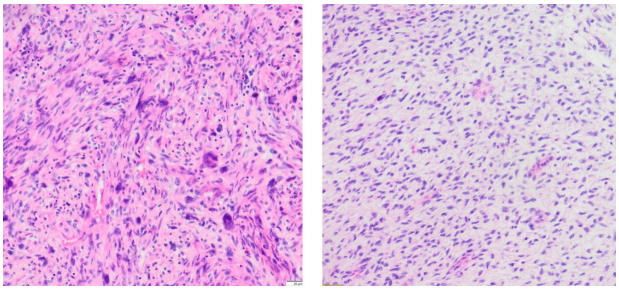

Figure 2. *Characteristic spindle neoplastic cells with pleomorphic nuclei (H/E)*

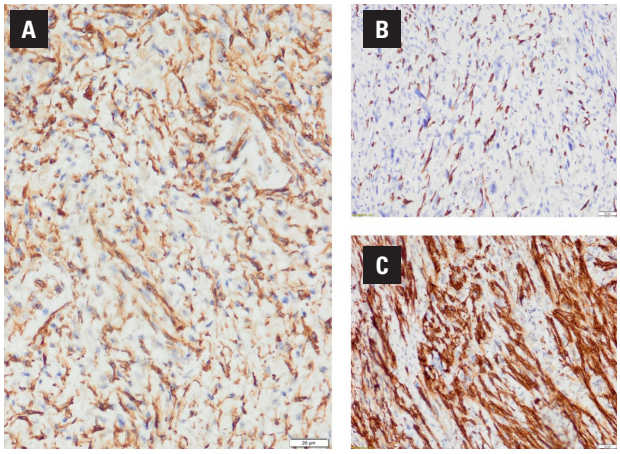

Figure 3. A. *Actin (+) IHC;* **B.** *Desmin (+) IHC;* **C.** *Caldesmon (+) IHC. Muscle cell differentiation is confirmed by positive for actin, desmin, and caldesmon staining. A focal positive desmin reaction is characteristic for smooth muscle tumours*

The diagnostic process of sarcomas located in the retroperitoneum is challenging due to its asymptomatic clinical picture, which was observed in other types of neoplasms as well [3]. Gradual growth of leiomyosarcoma will eventually apply pressure to adjacent structures, provoking abdominal pain, which might manifest as several other diseases. Thus, patients might report to the hospital with significant measures of the tumour. In the presented case, upper right abdominal pain was the only symptom of the tumour presence, which was impalpable.

In this case, primary adrenal leiomyosarcoma occurred in a patient with arterial hypertension and cardiac palpitation, which suggested pheochromocytoma. Due to the extreme rarity of leiomyosarcomas, they are not considered in differential diagnosis. Moreover, there are other malignant and benign adrenal tumours that might be found, such as non-functioning adenoma, myelolipoma, or adrenal cortical carcinoma, which is the most common malignant neoplasm [4, 5]. Therefore, plasma ACTH and cortisol examination together with dexamethasone suppression test are usually performed to confirm or exclude initial diagnosis. Adrenal biopsy is not advised due to optional adrenal crisis or tumour seeding. As a result, surgery remains a gold standard in the therapeutic conduct. There are still no characteristic markers of this malignancy, so diagnosis is based on the presence of neoplastic spindle cells in histopathological view positive for actin, desmin, and caldesmon. Consequently, ultimate leiomyosarcoma identification exclusively depends on the histopathological examination. The proliferation index (Ki-67) of the reported patient was estimated as 10–15%. Compared to other cases, the majority of tumours tested for Ki-67 were significantly higher [2, 4]. Identifying the tumour at such an early stage seems to be crucial to avoid infiltration of neoplastic cells to adjacent tissues and distant metastasis. Additional postoperative chemotherapeutics were not recommended on account of low Ki-67 and lack of surrounding organ infiltration and absence of metastasis. When needed, adjuvant therapy is generally composed of doxorubicin and dacarbazine [6]. In the case of patients with no recurrence or metastasis, periodic examinations are considered as an appropriate postoperative care.

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Conflict of interest
The authors declare that there is no conflict of interest.

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