CLINICAL IMAGE

Adrenocortical carcinoma with tumor thrombus extension into the right atrium

Krzysztof Szmyt[1], Tomasz Wierzbicki[1], Maciej Borejsza-Wysocki[1], Marek Jemielity[2], Sylwia Sławek-Szmyt[3], Łukasz Krokowicz[1]

1 Department of General, Endocrinological Surgery and Gastroenterological Oncology, Poznan University of Medical Sciences, Poznań, Poland
2 Department of Cardiac Surgery and Transplantology, Poznan University of Medical Sciences, Poznań, Poland
3 First Department of Cardiology, Poznan University of Medical Sciences, Poznań, Poland

A 34-year-old woman was admitted due to dyspepsia, nausea, and vomiting for several weeks. She lost 20 kilograms in the last 3 months. Her past medical history was irrelevant. Physical examination revealed a mass in the right upper quadrant of the abdomen and pitting edema of the lower extremities above the knee level. Comprehensive hormonal workup was within the normal range. Computed tomography (CT) demonstrated a right-sided enhancing retroperitoneal giant solid tumor (18 cm × 14.1 cm × 23.2 cm), with a regular lobed structure, extensive area of the central necrotic component, and high vascularity. The tumor mass modeled the adjacent organs with the right kidney displacement to the pelvis and compression of the right lobe of the liver (FIGURE 1A). There was no evidence of metastases or intra-abdominal or retroperitoneal lymphadenopathy. Moreover, a tumor thrombus extending into the inferior vena cava (IVC) and right atrium (RA) was detected (FIGURE 1B). Scintigraphy was negative for bone metastases. Based on these results, a clinical suspicion of nonsecreting adrenocortical carcinoma (ACC) was raised. The case was discussed by a multidisciplinary team and the patient was qualified for a one-stage procedure performed by a team of the general, vascular, and cardiothoracic surgeons. In the preparation for the surgery, endovascular embolization of the arteries supplying the tumor was performed by an interventional radiologist who administered polyvinyl alcohol particles measuring 500–700 μm and 700–1000 μm (FIGURE 1C). Subsequently, the patient underwent radical adrenalectomy via laparotomy with the Chevron incision along with midline sternotomy for simultaneous removal of intracaval and RA tumor thrombus invasion with the assistance of cardiopulmonary bypass and hypothermia (FIGURE 1D and 1E). Histopathologic examination confirmed the diagnosis

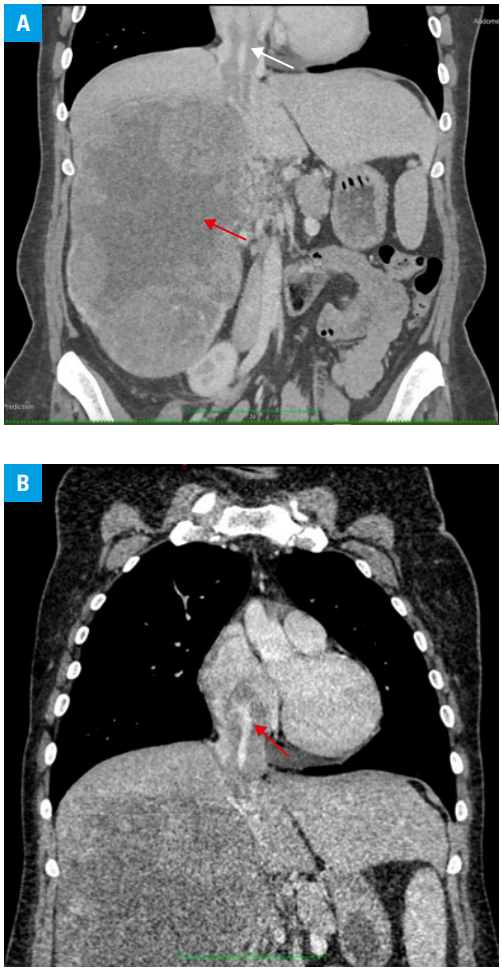

FIGURE 1 Adrenocortical carcinoma with venous tumor thrombus extension; **A** – contrast-enhanced computed tomography (CT) of the abdomen and pelvis showing a large tumor (18 cm × 14.1 cm × 23.2 cm), with tumor thrombus extending into the inferior vena cava; **B** – CT angiography of the chest showing the thrombus extending into the inferior vena cava and right atrium

Correspondence to:
Krzysztof Szmyt, MD, PhD, Department of General, Endocrinological Surgery and Gastroenterological Oncology, Poznan University of Medical Sciences, ul. Przybyszewskiego 49, 60-355 Poznań, Poland,
phone: +48 61 869 11 22,
email: krzysztof.szmyt@gmail.com
Received: December 30, 2022.
Revision accepted: February 1, 2023.
Published online: February 15, 2023.
Pol Arch Intern Med. 2023; 133 (3): 16434
doi:10.20452/pamw.16434

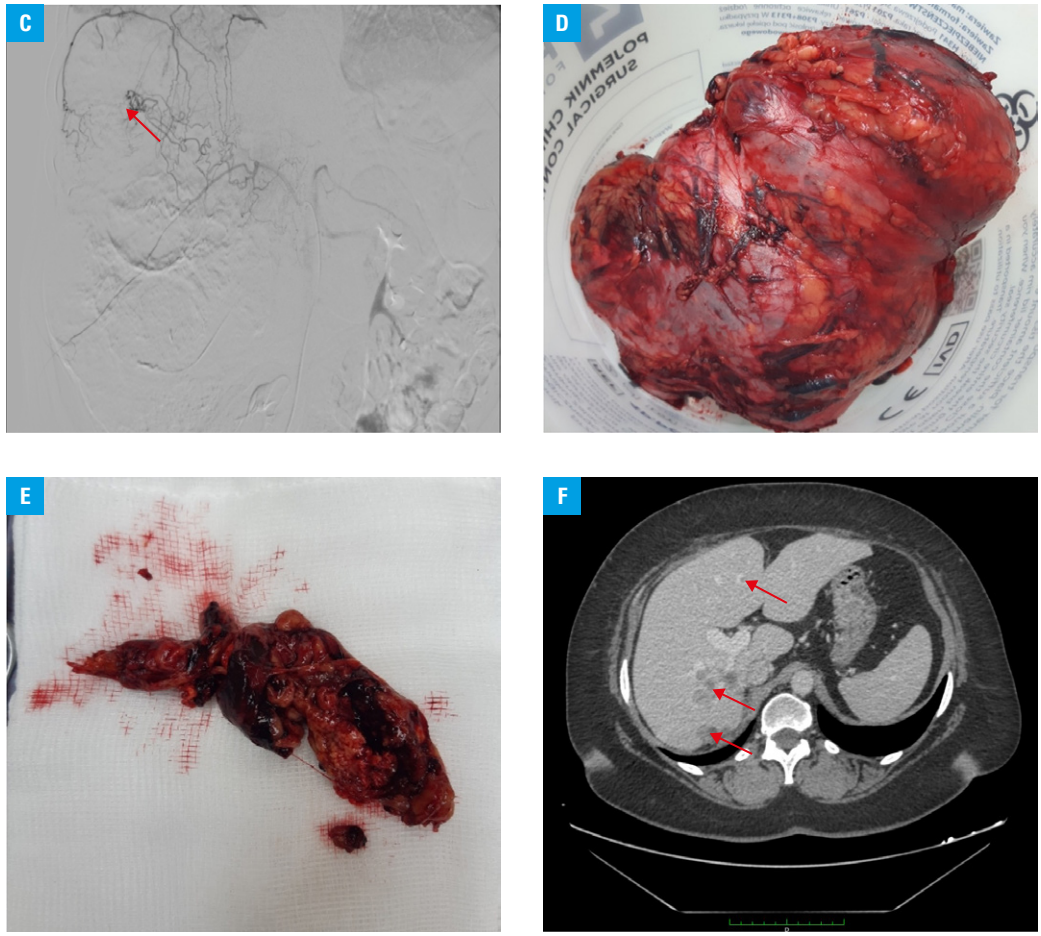

FIGURE 1 Adrenocortical carcinoma with venous tumor thrombus extension; **C** – endovascular embolization of the arteries supplying the tumor with polyvinyl alcohol particles; **D** – a complete right adrenal gland tumor specimen after the multidisciplinary surgery; **E** – the entire tumor thrombus specimen removed from the inferior vena cava and right atrium during the multidisciplinary surgery; **F** – CT 3 years after the surgery showing metastases in the lungs and liver

of ACC with tumor thrombosis of IVC and RA. The procedure and the postoperative course were uneventful. The patient was discharged on postoperative day 11 without any complications.

Three years later metastases were revealed in the lungs and liver (FIGURE 1F). The patient received a combination chemotherapy regimen with etoposide, doxorubicin, cisplatin, and mitotane. Finally, full regression of the disease was established on the chemotherapy treatment. Six-month follow-up was uneventful.

ACC is a rare and highly aggressive malignant tumor with a poor prognosis and estimated prevalence of 0.5 to 2 per million people.[1] More than 50% of ACC tumors present with a clinical / subclinical syndrome of hormone excess or manifestations related to tumor growth (mainly abdominal pain). However, hormonal evaluation is necessary to determine the secretory activity of the tumor. The gold standard for ACC staging is the chest and abdominal CT, but precise imaging evaluation of the tumor thrombus is essential to determine the surgical approach.[2] Although more extensive studies are needed to define optimal surgical treatment strategies in the cases of coexisting tumor thrombosis, radical (R0) resection (patients with localized disease) with histopathologic negative margins is the best option to achieve long-term survival rate and improve the quality of life.[3] In the previously reported ACC cases, the overall 5-year survival was still poor, at a level of approximately 40%.[4] Up to date, ACC with IVC and RA tumor thrombus requires highly specialized treatment in multidisciplinary clinical centers.[5]

ARTICLE INFORMATION

ACKNOWLEDGMENTS None.

FUNDING None.

CONFLICT OF INTEREST None declared.

HOW TO CITE Szmyt K, Wierzbicki T, Borejsza-Wysocki M, et al. Adrenocortical carcinoma with tumor thrombus extension into the right atrium. Pol Arch Intern Med. 2023; 133: 16434. doi:10.20452/pamw.16434

REFERENCES

1 Lafemina J, Brennan MF. Adrenocortical carcinoma: past, present, and future. J Surg Oncol. 2012; 5: 586-594.

2 Ciancio G, Farag A, Gonzalez J, Gaynor JJ. Adrenal tumors of different types with or without tumor thrombus invading the inferior vena cava: an evaluation of 33 cases. Surg Oncol. 2021; 37: 101544.

3 Verma Y, Mohith A, Shimjee S, et al. Adrenal tumor with inferior vena cava tumor thrombus: a case report and review of literature. J Endourol Case Rep. 2020; 6: 493-496.

4 Annamaria P, Silvia P, Bernardo C, et al. Adrenocortical carcinoma with inferior vena cava, left renal vein and right atrium tumor thrombus extension. Int J Surg Case Rep. 2015; 15: 137-139.

5 Fulawka L, Patrzalek D, Halon A. Adrenal cortical carcinoma with extension into the inferior vena cava - case report and literature review. Diagn Pathol. 2014; 9: 51.