

Primary Localized Giant Adrenal Cortical Carcinoma in a Young Adult Woman

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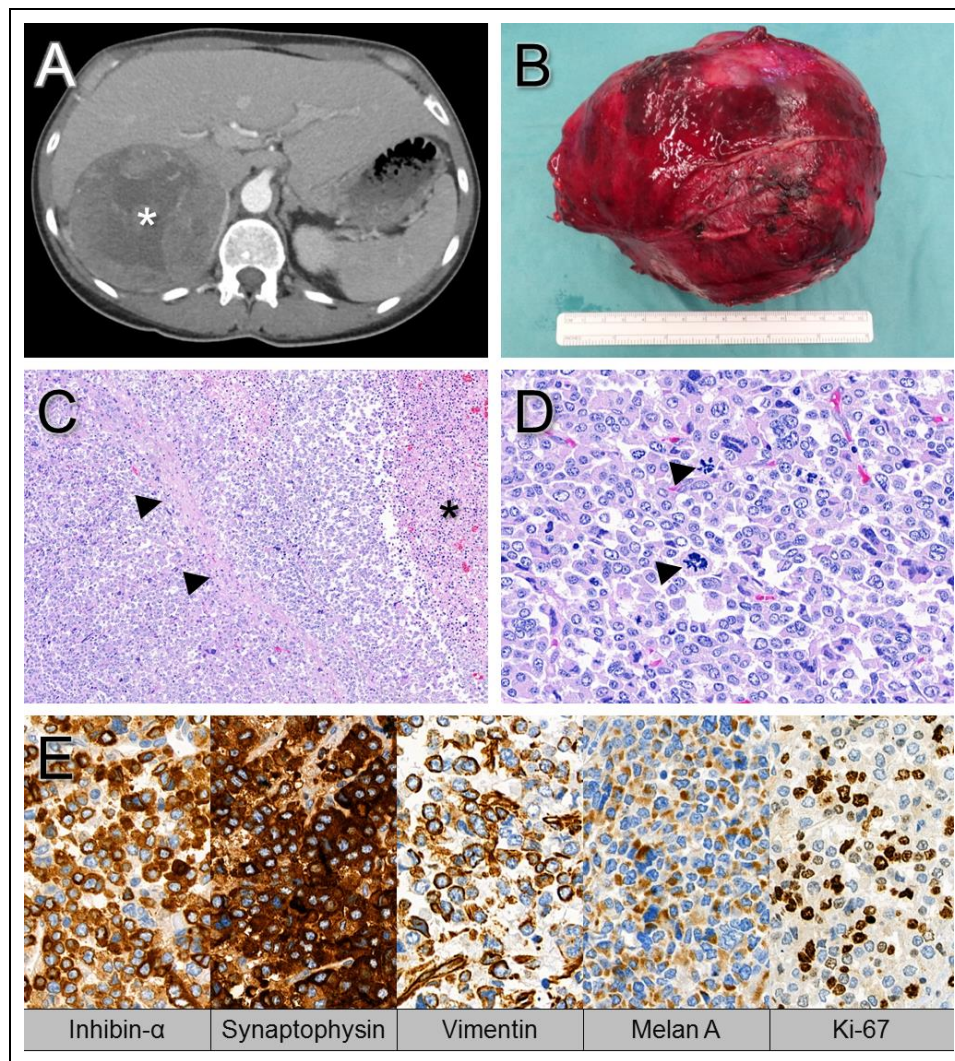


Figure 1 (A): Abdominal CT scans with 12 cm mass in the right adrenal gland, axial plane; **(B):** The enlarged adrenal gland (16 x 11 x 10 cm, 980 g; Figure 1B) with an encapsulated, multilobulated, partially necrotic tumor measuring 12 cm in diameter; **(C, D and E):** Histopathological examination of the adrenalectomy specimen. H and E stains (C, D) show a nodular proliferation of highly pleomorphic cells separated by fibrous bands (black arrowheads in C) and multiple areas of necrosis (black asterisk in A) with numerous mitoses (black arrowheads in D) and focal giant cell formation (black arrow in D). Immunohistochemical analysis (E) revealed strong and diffuse positivity for Inhibin- α and Synaptophysin, focal positivity for Vimentin and Melan A and an increased proliferation fraction (Ki-67 immunostain) of 40%. Magnification x100 in C, x400 in D and E.

A previously healthy 45-year-old woman was referred to our Department of Surgery with a three-month history of left flank pain, weight loss and intermittent fever associated with a large mass of the right adrenal gland discovered sonographically by the patient's general practitioner. Physical examination showed no abnormalities. Laboratory testing revealed an elevated C-Reactive Protein (CRP) level of 90 mg/L (<8 mg/L) and mild anemia (Hb 11.1g/L; 12-16g/L).

An unremarkable biochemical profile (aldosterone, cortisol, metanephrin, renin, sexual hormone) accompanied by constant normal clinical signs ruled out pheochromocytoma.

Abdominal Computed Tomography (CT) scans (Figure 1A) demonstrated a solid mass in the right adrenal gland with an average diameter of 12 cm. No distant metastases were found in the staging workup (stage II ACC).

Due to the patient's young age and the radiologic absence of metastatic disease our multidisciplinary team decided on surgical resection and the patient underwent a complete open right adrenalectomy (R0).

On gross examination, the enlarged adrenal gland (16 x 11 x 10 cm, 980 g; Figure 1B) showed an encapsulated, multilobulated, partially necrotic tumor measuring 12 cm in diameter. Microscopic examination revealed a nodular proliferation of highly pleomorphic cells separated by fibrous bands and multiple areas of necrosis with increased mitotic activity as well as capsular and lymphovascular invasion.

Immunohistochemical analysis revealed strong and diffuse positivity for Inhibin- α and Synaptophysin, focal positivity for Vimentin and Melan A and an increased proliferation fraction (Ki-67) of 40%. Based on morphology and immunohistochemistry, the diagnosis of an adrenal cortical carcinoma, high-grade, was established. The patient was discharged four days after surgery.

Considering the high Ki-67 expression, systemic adrenalitic chemotherapy with mitotane with curative intent for three years was recommended. Follow-up CT 12 months after surgery showed no signs of local recurrence.