Bladder paraganglioma. A case report

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Case Report

Urology

OPEN ACCESS

Background: Catecholamine-secreting tumors (CSTs) such as "pheochromocytomas" and "catecholamine-secreting paragangliomas" (CSPs) are derived from chromaffin cells of the adrenal medulla and sympathetic ganglia, respectively. Its prevalence is underestimated, since up to 50% of pheochromocytomas were diagnosed in autopsy series. Its distinction is important due to the implications for associated neoplasms, the risk of malignancy and genetic tests. We present the case of a 63-year-old female, referring an adverse event 18 years ago in a foley catheter placement presenting syncope and an adrenergic crisis. A year ago, an important weight loss was noted, an abdominal ultrasound was taken, showing evidence of a bladder tumor with presence of a bladder paraganglioma.

Keywords: Paraganglioma, pheochromocytoma, bladder tumor, hypertension, catecholamine-secreting tumor.

Catecholamine-secreting tumors (CSTs) such as "pheochromocytomas" and "catecholamine-secreting paragangliomas" (CSPs) are derived from chromaffin cells of the adrenal medulla and sympathetic ganglia, respectively (1). They are rare neoplasms, occurring in less than 0.2% of patients with hypertension. An annual incidence of approximately 0.8 per 100,000 person-years is estimated (2). Its prevalence is underestimated, since up to 50% of pheochromocytomas were diagnosed in autopsy series. It can occur at any age, more commonly between the fourth and fifth decade, with a 1:1 male and female ratio (3). CSTs are sporadic, but up to 40% have the pathology as part of a familial disorder, presenting with bilateral adrenal pheochromocytomas or paragangliomas (4). Generally diagnosed based on symptoms or an incidental discovery by imaging (60%). Its distinction is important due to the implications for associated neoplasms, the risk of malignancy and genetic tests. (5)

Case report

63-year-old female, referring an adverse event 18 years ago in a foley catheter placement presenting syncope and an adrenergic crisis. A year ago, an important weight loss was noted, an abdominal ultrasound was taken, showing evidence of a bladder tumor with presence of a bladder paraganglioma. In pelvic US, bladder tumor 44 x 47 x 36 mm. In abdominal CT a bladder mass in the anterior wall and roof of 43 x 21 x 64 mm with a volume of 31 cm3. Partial cystectomy of the 40 x 30 x 30 mm dome tumor was performed, reporting a paraganglioma, with lesion-free margins. Immunohistochemistry, GATA-3: +, Synaptophysin: +, S-100: +. Differential diagnoses such as stromal tumors bladder. Discharging the patient 48 hours after the surgical procedure, asymptomatic.

Discussion

The initial approach includes measurements of fractionated metanephrines in urine or plasma-free. All positive results require follow-up. Suggesting CT as initial imaging, but MRI is a better option in patients with metastatic disease or when radiation exposure must be limited. 123i-metaiodobenzylguanidine scintigraphy it is a useful imaging modality for metastatic CSPs. Patients with paraganglioma should be tested for succinate dehydrogenase gene mutations (SDHx) and those with metastatic disease for SDHB mutations. All patients with functional CSPs should undergo a preoperative block to avoid perioperative complications. Preparation should include a high-sodium diet and fluid intake to prevent postoperative hypotension. Minimally invasive adrenalectomy is recommended for most pheochromocytomas and open resection for most paragangliomas. International clinical practice guidelines suggest lifelong follow-up

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DOI 10.5281/zenodo.7444924

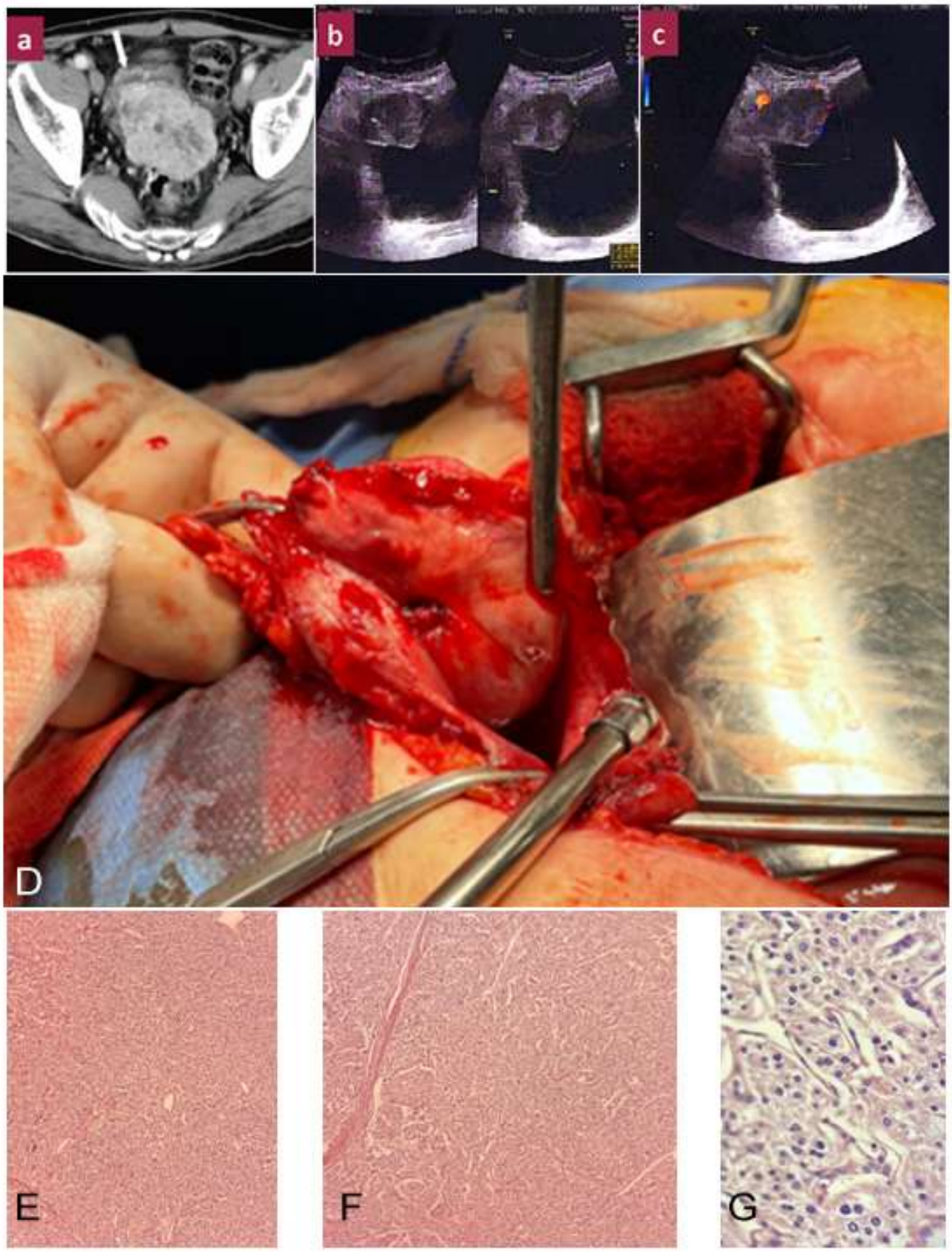

Figure 1. A. Contrasted CT scan. B. Bladder ultrasound. C. Bladder Doppler ultrasound. D. Domed bladder tumor (4x5 cm) resected. E, F, G. Microscopic description: Histological sections show an encapsulated neoplasm with a growth pattern in nests (Zellballen) surrounded by a prominent vascular network, it is made up of round to oval cells with abundant granular eosinophilic cytoplasm and nuclei with "salt and pepper" chromatin, focally with mild atypia characterized by increased nuclear size and hyperchromatism, without mitotic figures.

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to detect recurrent or metastatic disease. Bladder CSPs are rare tumors that can cause adrenergic symptoms.

Conclusion

The treatment of choice is surgical resection, with strict control of blood pressure, heart rate, and volume expansion. Personalized management with evaluation and treatment by multidisciplinary teams with adequate experience its necessary to ensure favorable results.

Conflicts of interests

The authors declare no conflict of interest

Acknowledgements

We deeply appreciate those who contributed to this case report.

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 DOI 10.5281/zenodo.7444924