

Functional medicine

Laparoscopic treatment of a cystic pheochromocytoma of 6cms: A challenging case



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ABSTRACT

Cystic pheochromocytoma is a very rare entity. Preoperative diagnosis is difficult because clinical, biochemical and radiologic findings are usually not consistent with a pheochromocytoma. Open surgery is traditionally the gold standard to avoid cyst rupture. We present a case of a 6 cm cystic pheochromocytoma treated by laparoscopy.

Background

Pheochromocytoma is a rare tumor. Its incidence is estimated of less than 0.1% in the global population. It usually develops from the medulla of the adrenal, but extra-adrenal pheochromocytoma can occur.

Although this tumor is usually solid, it can present as a cystic lesion.¹ Imaging and histology can differentiate between solid and cystic pheochromocytoma. The cystic form is due to hemorrhagic degeneration, necrosis, and cyst formation.¹ Few cases of cystic pheochromocytoma have been reported in the literature. We present a case of cystic pheochromocytoma that was challenging due to 4 factors: the size of the tumor more than 5 cm, the cystic form, severe symptomatology and difficulty to differentiate between benign and malignant form.

Case report

A 41 years old male with no medical history presented with intermittent colicky abdominal pain, episodes of palpitations and sweating for the past 2 years. On physical examination, he was found to have an elevated blood pressure of 170/90 mm Hg and no palpable abdominal mass. CT of abdomen showed 6 cm × 4.3 cm × 4.3cm cyst in the left adrenal gland with fine septations inside (Fig. 1). Laboratory investigations revealed a pheochromocytoma with plasma normetanephrine of 1087 pg/ml and metanephrine of 372 pg/ml. Serum potassium, plasma aldosterone, renin and urinary cortisol were normal. A case of cystic pheochromocytoma was suspected based on clinical,

biochemical and radiological findings. Laparoscopic transperitoneal left adrenalectomy was done (Fig. 2). During the whole surgery, there wasn't any significant hemodynamic instability. Surgical operation lasted 70 min. During post-op in-hospital stay he remained hemodynamically normal, with blood pressure around 120/70 mm Hg. No pain or surgical site infection have occurred. Patient was discharged home at 3rd postoperative day. Histopathology confirmed the diagnosis of a cystic pheochromocytoma with a PASS score of 3 (no metastatic potential) and negative margins (Fig. 3).

Discussion

Pheochromocytoma is a tumor arising from adrenomedullary chromaffin cells, which produce and secrete catecholamines. In the general population, the incidence of pheochromocytoma is about 0,005–0,1%.^{2,3} Cystic pheochromocytoma is even more rare with only few cases reported worldwide.³ The main clinical presentation of pheochromocytoma is hypertension, which is seen in 90% of cases, of which, two-thirds have paroxysmal hypertensive episodes, along with tachycardia, palpitations, headaches, sweating, tremors, and anxiety.¹ Pheochromocytoma should be considered as a diagnosis in patients presenting with nonspecific symptoms and an incidental adrenal mass. It can be difficult to diagnose cystic pheochromocytoma, as on imaging they can resemble to benign adrenal cysts and do not show the typical imaging features of solid pheochromocytoma. In our case, the patient was symptomatic with colicky abdominal pain, episodes of sweating

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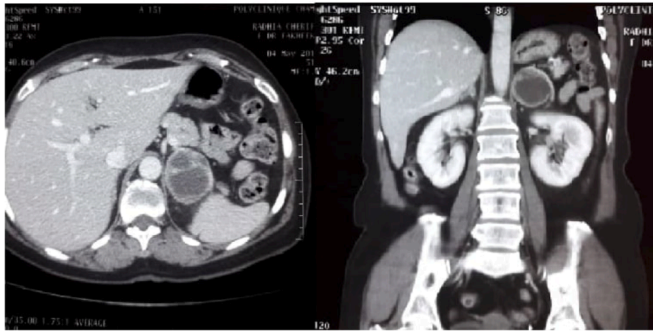


Fig. 1. CT scan showing a 6cm cyst in the left adrenal gland with fine septations inside.

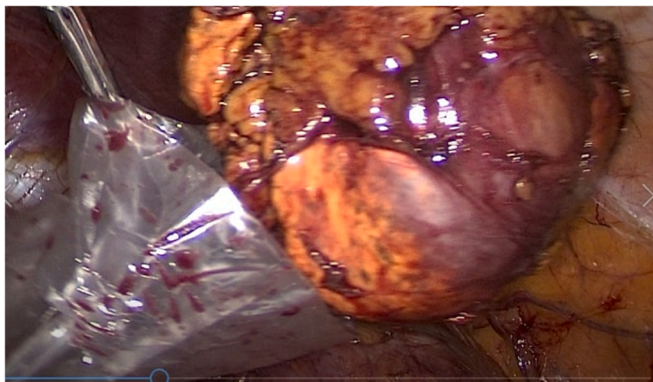


Fig. 2. Laparoscopic left adrenalectomy: Cystic mass being removed.

and palpitations. Increased plasma fractionated metanephrines made the diagnosis of a pheochromocytoma. Surgery is the curative treatment because, if left untreated, it can lead to several complications like

myocardial infarction, cardiac arrhythmias, heart failure, hypertensive encephalopathy, cerebro-vascular accident and sudden death.³ Therefore, blocking the effects of released catecholamines are recommended for all patients. laparoscopic surgery is effective and safe, traditional open surgery is the gold standard in the presence of adrenal tumors with suspicion of malignancy, like masses larger than 5 cm, because open surgery can increase R0 resection rate in case of malignancy.^{3,4} Minimal invasive techniques (laparoscopy and retroperitoneoscopy) have the advantages of less postoperative pain and ileus, less morbidity, improved cosmetics, and faster recovery, but with the negative impact in R0 resection and probably easier to cystic rupture occurrence.⁵ Our case was challenging because of the size (6cm), the cystic form and the severity of the symptoms. Thankfully, the patient was hemodynamically stable during the procedure and no rupture of the cyst has occurred. There are no studies comparing cystic pheochromocytoma rupture rates between minimally invasive surgery and open surgery. Comparisons have been made between transperitoneal laparoscopic method and the posterior retroperitoneoscopic approach. Lee et al.⁵ published a study where they concluded that retroperitoneoscopic adrenalectomy has shorter operative time, decreased abdominal viscera lesions because there is no incursion into the peritoneal cavity, less post-operative pain, and shorter time to first oral intake comparing to transperitoneal laparoscopic method. Direct access to adrenal gland is probably the reason for these advantages. In our case, transperitoneal surgery was performed due to the surgeon preference and the challenging factors listed above.

Conclusion

Cystic pheochromocytoma is rare and pose clinical, radiological and biochemical difficulties in making a correct preoperative diagnosis. Cystic rupture must be avoided when dealing with these lesions because of the risk of malignant cells spillage. To allow a secure R0 resection, open surgery is the gold standard. However, studies comparing open and minimally invasive techniques for large adrenal masses are lacking. In fact, few cases of cystic pheochromocytoma treated by posterior or transperitoneoscopic adrenalectomy were reported. More studies are needed to compare open and minimally invasive techniques in terms of resectability and cystic rupture rate.

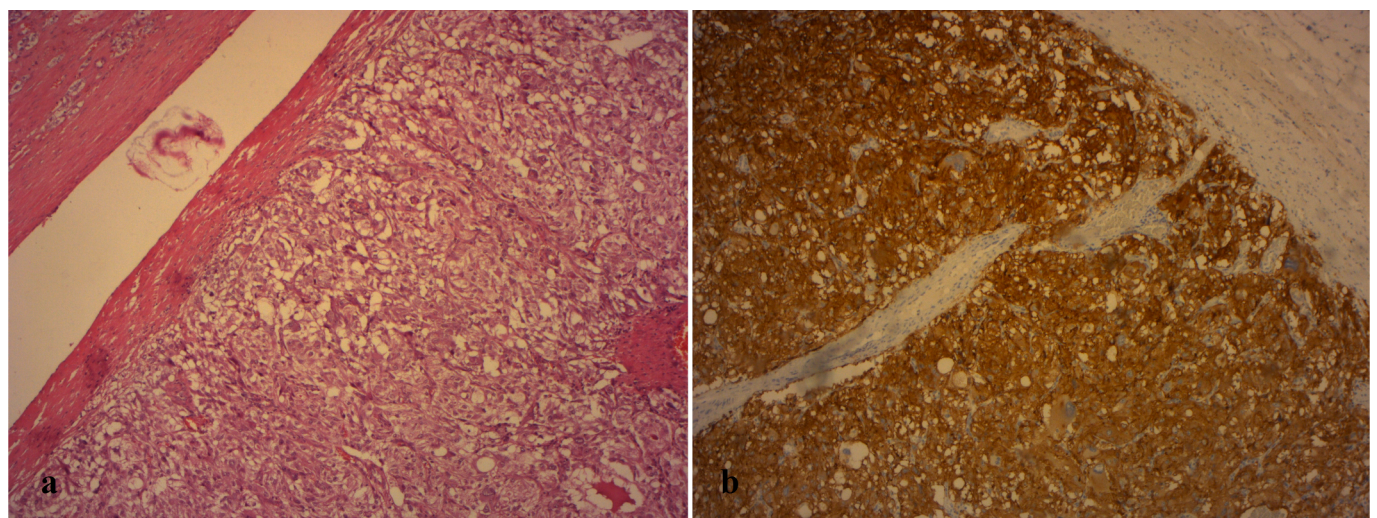


Fig. 3. a: A well encapsulated diffuse and dense proliferation of polygonal and large tumor Cells (HE x 50) b: Immunohistochemical study: tumor cells are positive for synaptophysin (synaptophysin x 50).

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