Case report

Simple adrenal cyst masquerading clinically silent giant cystic pheochromocytoma

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SUMMARY

Pheochromocytoma (PCC) may present as a cystic or solid tumour. Cystic PCCs are difficult to differentiate from simple cysts in the absence of classic symptoms of PCCs. Cystic nature develops due to intralesional bleeding and necrosis. We present a case of young man without any comorbidity who was diagnosed as a simple adrenal cyst and planned for laparoscopic excision but found to be functional PCC during the time of surgery only. The patient was managed with extensive monitoring and use of multiple drugs to control blood pressure. Surgery was completed without any complication laparoscopically and postoperative period was also uneventful. Histopathology confirmed the diagnosis of cystic PCC. Our case also shows the importance of functional imaging like metaiodobenzylguanidine (MIBG) scan in doubtful cases of adrenal cysts when other biochemical markers are unremarkable to diagnosis. We emphasise the importance of meticulous preparation for any intraoperative disasters even for apparently simple adrenal cyst.

BACKGROUND

Adrenal cyst is benign incidental tumours with about 300 cases reported in the literature.¹ With the widespread use of CT and MRI, the incidence of adrenal cyst has increased. The treatment depends on their nature (functional or non-functional), size and symptoms. Pheochromocytomas (PCCs) are rare catecholamine-secreting tumours originating from chromaffin cells of the adrenal medulla. They have an estimated incidence of 0.005%–0.1% in general population and 0.1%–0.2% among adult hypertensives.^{2,3} PCCs are usually solid tumours; however, cystic variants have been reported. Cystic PCCs are difficult to differentiate from the simple benign cyst in the absence of symptoms and structural resemblance in imaging. We report a case of 35-year-old male patient, diagnosed with large simple adrenal cyst taken up for laparoscopic excision, surprisingly was found to have PCC intraoperatively and confirmed on final histopathology report.

CASE PRESENTATION

A 35-year-old male patient with no previous comorbidities, presented with left back and flank pain of 2 months duration, which was moderate in intensity and relieved with oral analgesics. There was no history of headaches, palpitations and diaphoresis.

INVESTIGATIONS

Ultrasound abdomen reported a cystic septated mass of 13×8.5×8.6 cm in the left suprarenal region. The routine tests showed haemoglobin: 12.4 g/L, total leucocyte count: 5×10⁹/L, sodium: 134 mEq/L, potassium: 4.2 mEq/L, chloride: 110 mEq/L, serum urea: 32 mg/dL and creatinine: 0.9 mg/dL, and ECG was normal. A contrast-enhanced CT (CECT) abdomen showed large cystic mass 13.2×8.4×8.6 cm in the left adrenal gland. There was wall enhancement with thickening in peripheral aspect and intralesional thin septae. It was abutting tail and body of the pancreas with loss of fat planes ([figure 1A](#)). MRI T2 weighted image showed cystic lesion 13.2×8.4×8.6 cm with thick intermediated intensity walls and thin septae with mild peripheral thickening with no area of diffusion restriction ([figure 1B](#)). Echocardiography was normal. The biochemical test to detect catecholamine metabolites including 24 hours urinary vanillylmandelic acid (2.65 mg/24 hours), 24 hours urinary metanephrine's (120.5 mg/24 hours) and normetanephrine's (344.1 mg/24 hours) were all within normal limits. Plasma aldosterone was 10 ng/dL and aldosterone rennin ratio was 18, and within normal range. The serum cortisol was 4.2 ng/mL early morning sample. Further, 24 hours urinary cortisol was 24 mg which was within normal limits. Hydatid serology was negative.

DIFFERENTIAL DIAGNOSIS

Adrenal PCC.

TREATMENT

The patient was planned for laparoscopic adrenalectomy in view of symptomatic large adrenal mass. After confirming the fasting, patient was induced under general anaesthesia as per the institutional protocol. Intraoperative monitoring included ECG, pulse oximetry and invasive blood pressure. All the emergency drugs were kept ready for unforeseen intraoperative complications. Haemodynamics were well maintained throughout the induction until when the patient was positioned laterally for the surgery. After positioning, he developed first episode of hypertension (blood pressure (BP): 180/110 mm Hg) before pneumoperitoneum. This hypertension responded well to the intravenous nitro-glycerine (NTG) and sodium nitroprusside (SNP) infusion. Pneumoperitoneum was created and standard laparoscopy ports were placed as for adrenalectomy ([figure 2A](#)). The cystic tumour was

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Figure 1 (A) Contrast-enhanced CT abdomen coronal image showing homogenous cystic lesion with enhancing walls in left suprarenal area with thin septae. (B) MRI T2 weighted coronal image showing enhancing cystic lesion with enhancing walls arising from left adrenal gland.

seen bulging from the colonic mesentery. The descending colon and splenic flexure were mobilised medially (figure 2B). The Gerota's fascia between the ipsilateral kidney and adrenal was dissected and plane created between the two organs (figure 2C). During tumour dissection, BP rose up to 225/142 mm Hg with reflex bradycardia of pulse rate 49/min (figure 3). Patient had three to four similar episodes of hypertension and bradycardia during tumour resection. These episodes were managed by using NTG and SNP. The cystic tumour was dissected all around safeguarding the spleen, kidney and colon. A meticulous dissection was completed within an hour. After ligating adrenal vein, blood pressure came down to 90/40 mm Hg with pulse rate 110/min and pulse pressure variations of >25 mm Hg. The patient responded to 1 L of normal saline. Haemostasis was confirmed (figure 2D) and specimen delivered from left iliac region incision.

OUTCOME AND FOLLOW-UP

Postoperative events were uneventful. Cut specimen showed thick walled cyst with thin septae and areas of haemorrhage and

necrosis (figure 4A,B). Weight of tumour was 1100 g. Histopathology report confirmed diagnosis of PCC with cystic degeneration with Zellballen pattern. At 3 months follow-up, patient is doing well with no fresh complaints.

DISCUSSION

PCCs can present as cystic lesions which can mislead diagnosis. Giant PCCs are rare with male to female ratio 2:1.⁴ Classical symptoms include tachycardia, hypertension, sweating and headache which may not be present always. In a study by Pan *et al* on cystic PCC, 9 out of 15 cases were asymptomatic and in 6 out of 15 cases, diagnosis was suspected only at the time of resection due to variation in blood pressure, like in our case.⁴ One of the reasons for asymptomatic presentation in such cases is extensive necrosis causing significantly decreased catecholamine producing cells. Intraoperative complications among these cases included hypertensive crisis, hypotension, ectopic rhythm and cardiac arrest that responded to resuscitation and

Figure 3 The graphical representation of variations in the intraoperative haemodynamic parameters. The solid white arrows represent the episodes of significant bradycardia and hypertension.

Figure 2 (A) Intraoperative patient positioning in left flank up position with laparoscopy ports placed as for adrenalectomy. (B) Intraoperative image of descending and splenic flexure colon mobilised over adrenal cystic tumour. (C) Intraoperative image showing plane created between kidney and adrenal tumour by opening the Gerota's fascia. (D) Intraoperative image of tumour bed showing clipped adrenal vein and complete haemostasis.

almost all patient had cystic degeneration. In a series reported by Ito *et al* of 84 patients with PCC, 44% were partially cystic and 10% were highly cystic.⁵ It is recommended that in patients with adrenal cystic lesions with episodes of hypertension, 24 hours urinary catecholamines should be checked, which is raised in 95% of cases.⁶ Cystic PCC may not have classical symptoms and hence it is necessary to keep a high index of suspicion for PCCs. Cystic PCC may have very minimal functioning cells which produce and release catecholamines in paroxysmal manner. It is therefore difficult to diagnose such cases preoperatively with one time urinary and serum markers like metanephrine's. Failure to recognise them before resection can lead to intraoperative hypertension, postoperative hypotension and increased risk for cardiac events. Functional imaging like metaiodobenzylguanidine (MIBG) scan should be considered essential to prove the presence of sympatho-adrenal tissue⁷ as many times biochemical evaluation is not helpful and uptake in MIBG scan is definitive and aids in adequate preoperative preparation of such rare cases prior to surgical excision. Surgical approach in adrenal masses may vary depending on feasibility of mass excision. Large adrenal masses likely malignant carcinomas with possible infiltration into adjacent organs require open surgery and

Figure 4 (A) Gross specimen of the adrenal mass with soft cystic appearance. (B) Cut specimen showing thick walled cyst with areas of necrosis and haemorrhage.

multidisciplinary approach. Laparoscopy and robotics is done more frequently in view of being minimally invasive and speedy recovery. It requires minimal tissue handling, less blood loss, shorter hospital stay and less postoperative pain. Transperitoneal approach is favoured over retroperitoneoscopic because of large working space, direct access to adrenal vein and more favourable anatomy to the urologist.⁸ Costa Almieda *et al* reported a case of large adrenal cystic PCC operated by posterior retroperitoneoscopic approach and discussed its feasibility; however, narrow space and risk of rupture and spillage is the concern.⁹ Gupta *et al* reported a case of large cystic PCC operated by open approach and removed the mass with nephrectomy in view dense adhesion with renal hilum.¹⁰ Posterior retroperitoneoscopic approach is been frequently used now-a-days and results are comparable to transperitoneal approach. Advantages of this approach are direct access to adrenal gland being a retroperitoneal organ, no breach of peritoneum, less blood loss, shorter hospital stay and less post-operative pain. A meta-analysis conducted by Young *et al* which included eight full length articles for comparing transperitoneal versus retroperitoneal laparoscopic adrenalectomy reported that retroperitoneal approach may be superior to transperitoneal

approach due to short operative time and shorter hospital stay. However, it was also mentioned that retroperitoneal approach requires a significant learning curve and that it's a difficult procedure in obese patients.¹¹ Chung *et al* reported the feasibility of laparoscopic adrenalectomy using retroperitoneal approach for large size PCC and outcomes were comparable.¹² Both the techniques are similar and outcomes are comparable in the hands of experienced laparoscopic experts in field of urology.

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Patient's perspective

I am very thankful to the whole team of doctors for handling my case very well and managed it.

Learning points

- Giant cystic pheochromocytomas are very rare.
- Typical classical symptoms may not be present in view of minimal functioning cells.
- High index of suspicion should be kept and investigate properly before taking the patient for surgery.
- MIBG scan is an useful test in such cases to diagnose the functioning adrenal masses.
- Intraoperative anaesthetic management is very crucial for controlling hypertension and preventing any cardiac events.

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