CASE REPORT

Adrenal ganglioneuroma in a patient with polycystic ovarian disease (PCOD): a rare association

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SUMMARY

Adrenal ganglioneuromas are rare, benign incidentalomas of a neural crest origin. A majority of these tumours are clinically silent and discovered on imaging for unrelated reasons. Polycystic ovarian disease (PCOD) is an endocrine disorder characterised by bilateral polycystic ovaries, anovulation leading to infertility, irregular menstrual cycles and features of androgen hormone excess. Herein we report a rare case of adrenal ganglioneuroma in a 14-year-old girl with PCOD. She was referred to us by the gynaecologist after incidental detection of adrenal mass on ultrasonography. Except for raised 24 h urinary metanephrines, rest of the hormones measured were in normal range. Transperitoneal adrenalectomy was performed and histopathology was suggestive of ganglioneuroma. Postoperative recovery was excellent and she is doing well. To our knowledge it is the first such type of case to be reported.

BACKGROUND

Ganglioneuromas (GNs) are rare benign tumours of the sympathetic nervous system chain (originating from the primordial neural crest cells) and are commonly located in the posterior mediastinum and retroperitoneum. They are rarely found in the adrenal gland and only few cases have been reported until now. Adrenal incidentalomas are found in 1–10% of abdominal CT scans and of these 1–6% are GNs.¹ Most cases have been reported in children or young adults after incidental detection on imaging. They are often silent and rarely produce sympathetic amines and symptoms of sympathetic overactivity. Association of polycystic ovarian disease (PCOD) with adrenal GN is rare and no such cases have been reported until now.

CASE PRESENTATION

A 14-year-old girl presented with a history of irregular menses and dull aching, intermittent left flank pain for the past 6 months. There was no history of a similar illness in any family member. On clinical examination the patient was normotensive without any palpable mass in the flank region.

INVESTIGATIONS

At 24 h urinary metanephrines—2.17 mg (normal 0–1 mg/24 h). Serum sodium 145 meq/L and serum potassium 4.2 meq/L.

Ultrasonography was suggestive of bilateral PCOD with a left adrenal mass. Contrast-enhanced CT showed a 10×9 cm, heterogeneously enhancing, non-calcified left adrenal mass with areas of

necrosis (figure 1). CT-guided fine-needle aspiration cytology was suggestive of adrenocortical adenoma.

DIFFERENTIAL DIAGNOSIS

Adrenal GNs should be included in differential diagnosis of any adrenal or retroperitoneal mass.

TREATMENT

She underwent transperitoneal adrenalectomy by a left subcostal incision (figure 2) under general anaesthesia. Intraoperatively there was mild tachycardia with normal blood pressure. The adrenal mass was lying over the hilum and upper pole of the left kidney and was also displacing the tail of pancreas anteriorly.

OUTCOME AND FOLLOW-UP

Postoperative recovery was excellent and she was discharged on day 5. Histopathological examination was suggestive of adrenal GN (figure 3). The patient is doing well at 1 year of follow-up.

DISCUSSION

Association of adrenal GN with PCOD is a rare occurrence. GNs are rare incidentalomas of the neural crest origin. Neuroblastoma, pheochromocytoma and ganglioneuroblastoma are some other neural crest derived tumours. Adrenal GNs mainly affect young adults without sex predominance, cases have also been reported in children.^{1–3} Although often clinically silent, GNs occasionally are symptomatic due to catecholamines or androgen production.^{4–5} They can very rarely metastasise to distant sites.⁶ Characteristic CT findings include non-enhanced low attenuation <40 HU, punctuate calcifications; MRI findings include low-signal

Figure 1 Contrast-enhanced CT of the kidney, ureter and bladder showing a left adrenal mass pressing the hilum and upper pole of the left kidney.

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Figure 2 Intraoperative images. (A) Left renal vein (white arrow). Left adrenal vein (yellow arrow). (B) The excised adrenal mass.

Figure 3 Histopathology demonstrates mature ganglion cells (black arrow) set in an abundant mixture of mature Schwann cells (red arrow).

Learning points

- ▶ Adrenal ganglioneuromas are benign incidentalomas originating from the primordial neural crest cells.
- ▶ Occasionally the tumour may be symptomatic due to elaboration of hormones or due to mass-related effects.
- ▶ A CT scan and MRI are helpful and may reveal characteristic features.
- ▶ Treatment may be by laparoscopic adrenalectomy or open adrenalectomy.
- ▶ Histopathology of resected tumour gives a definitive diagnosis.

intensity on T1-weighted images and high-signal intensity on T2-weighted images.^{5–7} Treatment may be by open or laparoscopic adrenalectomy.⁴ Patients with PCOD present with features of androgen excess produced by ovary and sometimes by adrenal androgens due to an adenoma. So in a patient with PCOD presenting with a simultaneous adrenal mass, GN should also be a differential diagnosis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Commissioned; internally peer reviewed.

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