Development of a large adrenal myelolipoma in the context of long-term elevated ACTH

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DESCRIPTION

Adrenal myelolipomas are the second most common benign tumours of the adrenal gland. Despite their frequent occurrence, there is uncertainty around their aetiology. We present the case of a giant adrenal myelolipoma that supports the role of prolonged adrenocorticotrophic hormone (ACTH) stimulation in the induction of myelolipomas in congenital adrenal hyperplasia (CAH).

A 27-year-old man with salt-losing CAH due to 21 hydroxylase deficiency was initially reviewed in the endocrine department in 1996. He was taking a total of 0.75 mg of dexamethasone daily—0.5 mg in the morning and 0.25 mg at night. He clinically appeared cushingoid with abdominal striae, centripetal obesity and hypertension. A CT scan at that time showed normal adrenal glands (figure 1, panel B). Dexamethasone was switched to prednisolone and then later to hydrocortisone to reduce the side effects of excess glucocorticoid exposure. As a consequence, his ACTH and 17-hydroxyprogesterone (17-OHP) levels were persistently elevated from the age of 27 years to the present day.

In 2018, at the age of 50 years, he presented to the emergency department with severe abdominal pain and a CT scan revealed a 15×16×19 cm mass in the left adrenal gland. Radiology review reported features of a giant adrenal myelolipoma (panel A). At this time, his morning plasma ACTH was high at 486 ng/L (reference range 7.2–63.3) and 17-OHP levels were 3337 ng/dL (reference range <198 ng/dL). He underwent laparoscopic excision of 2030 g adrenal myelolipoma (macroscopic and microscopic appearances shown in panels C and D).

Adrenal myelolipomas consist of fat and haematopoietic tissues and are hormonally inactive. They are usually asymptomatic but can present with abdominal pain, spontaneous rupture and retroperitoneal haemorrhage. There are no guidelines for treatment but for large, rapidly growing tumours and those with compressive symptoms, surgery is considered. Up to 86% of myelolipomas are detected incidentally on imaging,¹ and the incidence is expected to rise with the increased use of abdominal imaging in clinical practice.

The aetiology of myelolipomas is not established, but the increased incidence in patients with CAH and Cushing's disease, both of whom have raised ACTH levels, has suggested a role for prolonged ACTH stimulation.¹ The role of hormones in inducing adrenal tumours was first proposed by Selye and Stone in 1950 who was able to induce bone marrow tissue like changes by injecting anterior pituitary hormone

Figure 1 (A) CT scan of abdomen (2018) showing left giant adrenal myelolipoma (arrow), (B) CT scan (1996) of abdomen showing normal left adrenal gland (arrow), (C) macroscopic appearance of tumour and (D) histology of left adrenal cortex showing fat and haematopoietic tissue.

Learning points

- ▶ Adrenal myelolipomas are the second most common benign tumours of the adrenal gland, and their incidence is expected to rise with the increased use of abdominal imaging in clinical practice.
- ▶ Adrenocorticotrophic hormone may play an important role in induction of these tumours.

extracts into normal rat adrenal glands.² The increased expression of *Melanocortin two receptor (MC2R)* on adrenal myelolipomas further supports the role of ACTH.³ Prolonged exposure to high ACTH levels for over two decades in association with a large myelolipoma in this patient provides further evidence for the role of ACTH in the induction of myelolipomas.

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