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ENDOCRINE IMAGE

Primary Pigmented Nodular Adrenocortical Disease: A Rare Cause of Cushing's Syndrome

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ABSTRACT

Primary pigmented nodular adrenocortical disease (PPNAD) is one of the rare cause of adrenocorticotropic hormone (ACTH) independent Cushing's syndrome. More than 90% of the reported PPNAD have been associated with Carney's complex. Primary pigmented nodular adrenocortical disease is one of the major criteria for the diagnosis of Carney's complex (CNC). We report a case of PPNAD which is not associated with CNC.

Keywords: Cushing's syndrome, Primary pigmented nodular adrenocortical disease, Bilateral adrenalectomy.

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INTRODUCTION

Primary pigmented nodular adrenocortical disease (PPNAD) is a rare cause of adrenocorticotropic hormone (ACTH) independent Cushing's syndrome. It can be associated with Carney's complex (CNC) and has a characteristic gross and microscopic appearance. Autopsy reports suggest almost every case of CNC is associated with PPNAD. All PPNAD is associated with Cushing's syndrome with overt manifestations in 70% of cases.

CASE REPORT

We intent to present a 15-year-old girl who presented with classical Cushingoid symptoms with multiple pigmented spots over her trunk. Biochemical evaluation confirmed ACTH independent Cushing's syndrome. Contrast enhanced computed tomography (CECT) of the abdomen revealed bilateral heterogeneously enhancing hypodense adrenal lesions with washout in delayed scans (right adrenal 2.6 × 1.7 cm (HU 37) and left adrenal 2.1 × 1.7 cm (HU 12). Further evaluation did not reveal other components of CNC. After adequate preoperative preparation, she underwent bilateral laparoscopic adrenalectomy. Macroscopically, both the tumors were bright yellow and, on cut section, there were blackish discoloration which was more evident on the right side (Fig. 1). Microscopy confirmed the diagnosis of primary pigmented nodular adrenal hyperplasia.

Primary pigmented nodular adrenocortical disease is seen in 25 to 40% of CNC patients. However, histological evidence of PPNAD has been reported in almost every patient with CNC.[1] Primary pigmented nodular adrenocortical disease usually presents in the 2nd or 3rd decade of life with no sex predilection.[4] The hypercortisolism is resistant to dexamethasone suppression test. Contrast enhanced computed tomography may show normal or pigmented nodules as string of pearls appearance or round, well delineated and hypodense lesions of both adrenals.[2] Bilateral adrenalectomy is indicated to avoid the morbidity and mortality associated with CS. The gross and histological appearance is very characteristic of PPNAD. Macroscopically, adrenal glands are usually normal or slightly enlarged in size; on cut section shows multiple dark brown or black nodules ranging from 0.5 to 5 mm. Microscopically, numerous nonencapsulated cortical nodules containing large amount of lipofuscin or neuromelanin, responsible for their black appearance and a pathognomonic atrophy

Fig. 1: Cut section of both adrenals displaying the characteristic brown black pigmentation on the right adrenalectomy specimen

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of the remaining cortex is usually observed.[3] Molecular studies have demonstrated inactivating mutations of the PRKAR1A gene on chromosome 17q22-23 or of the PDE11A gene.[4] Lifelong follow-up and screening with annual echocardiogram, thyroid and testicular ultrasound is recommended to detect other components of CNC.[5]

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