Aldosterone-secreting adrenocortical adenoma in children

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ABSTRACT

Aldosterone-secreting adrenocortical adenoma is an unusual cause of secondary hypertension in childhood. We describe an eight year-old female child who presented clinically with weakness of lower limbs, headache and hypertension. Thorough clinical, laboratory and radiological evaluations confirmed the diagnosis of hyperaldosteronism due to a left adrenocortical adenoma. Left adrenalectomy after preoperative stabilization resulted in satisfactory recovery. Histopathological examination confirmed the diagnosis of an aldosteronoma. We review our experience with this rare entity and the pertinent literature.

KEY WORDS: Aldosteronoma, primary hyperaldosteronism

INTRODUCTION

Aldosterone-producing adrenocortical adenoma remains an extremely rare entity in the pediatric age group. To the best of our knowledge, only a few cases have been reported in (English) literature.[1-4] It is an uncommon cause of secondary hypertension in children and its accurate diagnosis demands recognition of its typical clinical presentation so that an unduly high morbidity associated with delayed diagnosis and resulting complications can be avoided by appropriately early surgical intervention. Thus, the reporting of the present case seeks to make physicians aware of clinical presentation and the management protocol of this rare entity.

CASE REPORT

An eight year-old female child presented with symptoms of headache and with aching pain and discomfort in both lower limbs that gradually resulted in difficulty in walking over a period of 15 days. The child gave a history of occasional episodes of palpitation and dizziness for the last one month. There were no other associated respiratory, cardiovascular or neurological symptoms.

On examination, the child appeared active, moderately nourished and anemic with a pulse rate of 86/minute, blood pressure of 156/96 mm Hg (right upper limb in supine position) and a normal body temperature. Neurological examination of the lower limbs suggested muscular weakness was present in both legs with a motor power of 3/5. The cardiovascular, respiratory and abdominal examinations revealed no abnormality.

Laboratory investigations showed hemoglobin of 8.8%, normal total and differential leukocyte counts and normal liver function tests. The evaluation of renal function showed severe hypokalemia with serum potassium levels of 2.8 mEq/dl and normal blood urea, serum creatinine and serum electrolyte levels. The assessment of metabolic status of the patient showed the presence of metabolic alkalosis (pH 7.44, partial pressure of carbon dioxide (PaCO₂) of 47 and serum bicarbonate 30 mEq/L). Ultrasound study of the abdomen suggested the presence of a well-circumscribed left adrenal mass measuring 3 cm x 2.4 cm with a normal right adrenal gland; both kidneys were normal [Figure 1]. This provided a clue to the diagnosis and an elevated serum aldosterone level of 2.5 nmol/l (normal: 0.1-
0.97 nmol/l in the presence of normal cortisol levels confirmed the diagnosis of primary hyperaldosteronism. The 24 h excretion of vanillylmandelic acid (VMA), norepinephrine and epinephrine after correction of hypokalemia, was also normal.

Clinical, laboratory and radiological evaluations confirmed a diagnosis of primary hyperaldosteronism due to an aldosterone-secreting adenoma and surgical intervention was planned after preoperative stabilization of the physiological status of the patient. Intravenous potassium infusion under electrocardiographic monitoring followed by oral potassium supplementation resulted in initial correction of the severe hypokalemia and subsequent maintenance of normal potassium levels. Muscular weakness disappeared after correction of hypokalemia. After achieving a normotensive state with oral spironolactone, surgical exploration was performed. A well-circumscribed, encapsulated left adrenal mass measuring approximately 3 cm in diameter was found with normal vascularity of the tumor bed. Both kidneys and the right adrenal gland appeared normal on gross examination. A left adrenalectomy was performed. Histopathological examination of the resected specimen showed adenoma cells arranged in cords and trabeculae with an absence of areas of hemorrhage, necrosis, atypical mitosis or vascular invasion [Figure 2]. The postoperative period remained uneventful and the patient recovered satisfactorily. Serum aldosterone levels became normal and the antihypertensive drugs were tapered over a period of two weeks. An ultrasound study performed six months after surgery revealed no abnormality. The patient has been followed up for eight months and is doing well.

DISCUSSION
Aldosterone-producing adrenocortical adenoma is an extremely uncommon cause of secondary hypertension in the pediatric age group.\(^1,5\) Although bilateral adrenal hyperplasia has been reported in etiology of primary hyperaldosteronism and hypertension in childhood, surprisingly few aldosterone-secreting adrenal adenomas have been reported. We are aware of only 19 cases that have been reported in (English) Literature.\(^1,3-5,7\)

Histopathologically, most of the aldosterone-secreting adrenocortical adenomas are solitary, well-circumscribed benign lesions measuring < 2 cm in size without any evidence of hemorrhage, necrosis and any local or distant metastasis.\(^1\) Although malignant tumors are very rare, they still need to be considered and differentiation is necessary for prognosis.\(^1\) An objective assessment by Weiss criteria appears very useful for assessing the malignant potential of the tumor.\(^1,8\) The presence of three or more of the following nine features as proposed by Weiss et al. predicts the malignant behaviour of adrenocortical adenomas: nuclear grade III or IV, atypical mitoses, clear cells constituting less than 25% of the tumor, mitotic rate greater than 5 per 50 high-power fields, diffuse architecture, necrosis, invasion of venous and sinusoidal structures and invasion of capsule.\(^1,8\) Although the size of the adenoma reported here was > 2 cm, the absence of the above features suggested the benign nature of the adenoma.

The clinical features in the present case are typical of those described by most investigators in literature. According to our and others' experiences, the clinical presentation is usually heralded by features of headache, palpitation, dizziness due to presence of hypertension and features of weakness, discomfort, aching pains in the lower limbs which occurs due to metabolic abnormality, mainly hypokalemia.\(^1-7\)

A thorough clinical laboratory and radiological evaluation usually confirms the diagnosis of aldosteronoma. The presence of hyperaldosteronism with normal cortisol levels usually provides a clue to diagnosis.\(^1,9\) In doubtful cases, either bilateral adrenal vein sampling for aldosterone estimation or presence of unsuppressed high serum aldosterone levels in the presence of low basal and stimulated renin activity confirms the diagnosis and even helps in localization of the lesion.\(^1,9-11\) Ultrasound scanning has been found to be of little help in the diagnosis of adrenal adenomas, mainly due to the small size of the tumor. However, as in the present case and a few reported cases, it is useful in the presence of a large adenoma.\(^1\) In doubtful cases, computed tomography (CT) scan, magnetic resonance imaging (MRI) and adrenal scan with iodine \(^131\)-19-Iodocholesterol have been recommended for localization of the tumor, to exclude contralateral involvement and to detect multiple adenomas.\(^1,2,5,6,9,10\)

The management of an aldosterone-secreting, adrenocortical adenoma aims at complete surgical excision after preoperative stabilization of physiological

**Figure 2**: Photomicrograph showing adenoma cells arranged in cords and trabecular with a well-defined fibrous capsule with an absence of mitosis (H/E, Light microscope, 40x)
status of the patient. Adrenalectomy remains the surgical procedure of choice in such cases. As seen here and in other cases, oral spironolactone (2-4 mg/kg/day) along with oral potassium supplementation helps in control of the hypertension and correction of the hypokalemia, thus resulting in stabilization of the deranged physiological status in the preoperative period. Moreover, preoperative drug therapy facilitates the recovery of the suppressed contralateral zona glomerulosa thus preventing postoperative hypoaldosteronism. A similar surgical approach resulted in a satisfactory recovery in the present and other reported cases.

We conclude that although rare, aldosterone-secreting adrenocortical adenoma should be considered as a differential diagnosis in children presenting with secondary hypertension. Undue high morbidity associated with delayed diagnosis and resulting complications can be avoided by appropriate early surgical intervention.

REFERENCES


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