Reninoma presenting as cardiac syncope

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ABSTRACT

Reninoma, a renin-secreting tumor of the juxta-glomerular cells of the kidney, is a rare but surgically treatable cause of secondary hypertension in children. We report a case of reninoma presenting as cardiac syncope with long QTc on electrocardiogram due to hypokalemia.

Keywords: Hypokalemia, reninoma, secondary hypertension

INTRODUCTION

Reninoma causes hypertension and hypokalemia via hypersecretion of renin. We describe a case of reninoma and provide a review of the literature. The subject presented with syncope and had hypokalemia, long QTc and hypertension with concentric left ventricular hypertrophy. Imaging studies revealed the presence of a lesion in the renal cortex, which, on immunohistochemistry, was identified as a juxta-glomerular cell tumor (Reninoma). Following nephrectomy, the blood pressure and hypokalemia normalized.

CASE REPORT

An 8-year-old, thin, lean boy presented with a history of two episodes of syncope. On examination, pulse rate was 110 beats per minute and the blood pressure was 210/110 mmHg. There was no radio-femoral delay. A heaving apex in the fifth intercostal space inside the mid-clavicular line, a wide, but not-fixed split S2 with normal P2 and a grade II/VI systolic murmur at the apex were noted. On abdominal examination, a renal bruit was heard on the left side. Neurological examination and fundus were normal. His electrocardiogram revealed sinus tachycardia with corrected QT interval of 520 msec and occasional premature ventricular complexes [Figures 1 and 2]. There was no family history of deafness or sudden death.

Laboratory studies were remarkable for serum sodium 136 mEq/L, potassium 2.1 mEq/L, chloride 99 mEq/L, bicarbonate 31 mEq/L, blood urea 22 mg/dL and serum creatinine 0.8 mg/dL. Serum calcium, 24-h urinary vanillyl mandelic acid (VMA) levels and complete blood counts were within normal limits. Results of the urinalysis were normal.

Echocardiography revealed concentric left ventricular hypertrophy and mild mitral regurgitation. There was no evidence of left ventricular outflow tract obstructive disease. The patient initially received intravenous magnesium and potassium, nitroglycerine, oral beta blockers and angiotensin receptor antagonists. His QTc normalized, premature ventricular ectopics disappeared, blood pressure got controlled and the patient became asymptomatic. Renal ultrasound Doppler was suggestive of replacement of left kidney by a necrotic mass (5 cm × 4.8 cm), with invasion of the left renal artery with normal vascular flow profile. A contrast enhanced computed tomography scan of the abdomen confirmed a cystic mass replacing the left kidney and compressing the left renal vein, with normal adrenal glands. A differential diagnosis of Bosnaik type IV cyst and Wilm's tumor were considered [Figure 3].

The patient was referred for resection of the left renal mass. Nephrectomy was performed and a histopathological examination of the specimen revealed features suggestive of reninoma [Figure 4]. The post-operative period remained uneventful. Patient's blood pressure became normal. His symptoms resolved and he is asymptomatic on one and half years follow up.

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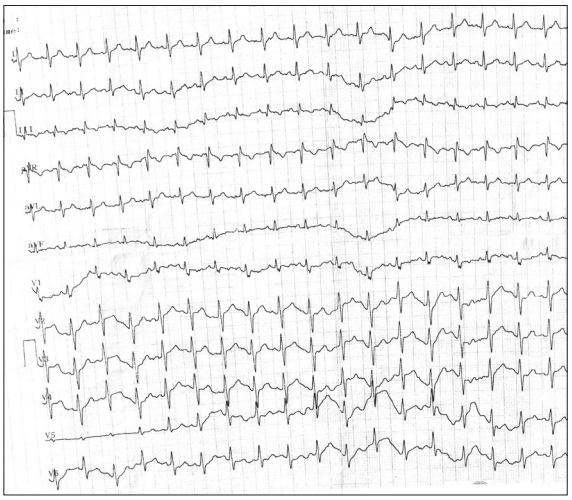

Figure 1: Electrocardiogram of the patient showing sinus tachycardia and a prolonged QTc

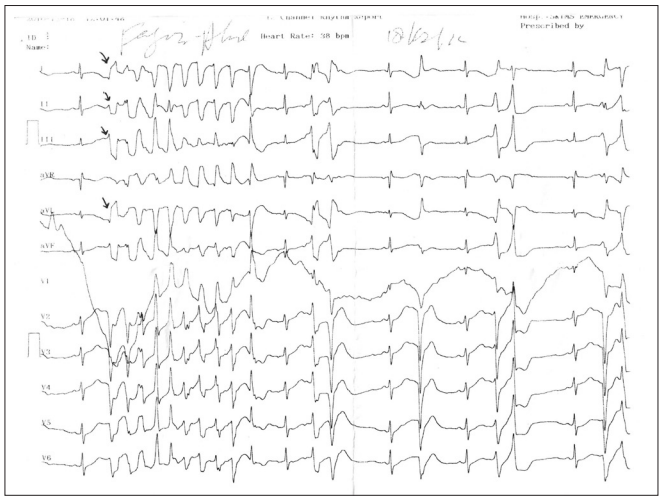

Figure 2: Electrocardiogram of the patient showing frequent ventricular ectopics and a run of nonsustained ventricular tachycardia

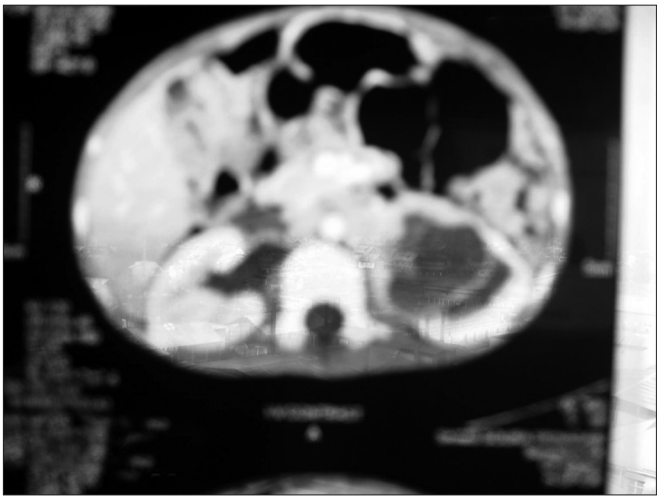

Figure 3: Computed tomography picture showing renal mass lesion

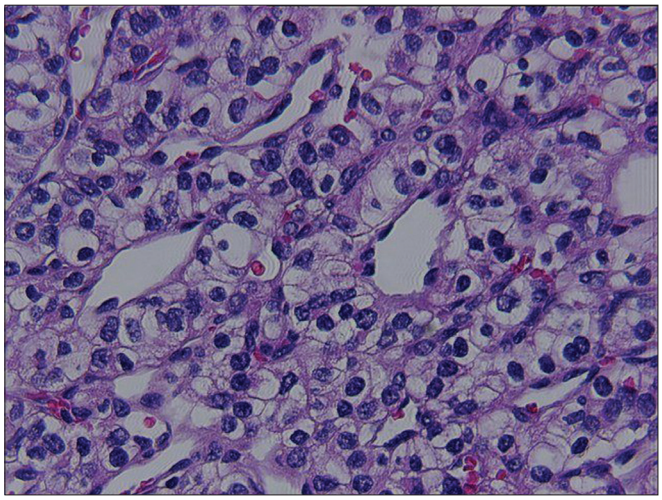

Figure 4: Histopathology consistent with Reninoma

DISCUSSION

The prevalence of hypertension in children ranges from 1% to 5%. Secondary causes of hypertension are more common in children than in adults.[1] Mineralocorticoid excess should be considered in patients presenting with the triad of hypertension, unexplained hypokalemia and metabolic alkalosis.[2] Primary aldosteronism caused by aldosterone-secreting tumors or idiopathic adrenal hyperplasia is rare in childhood, but has been described.[3] Excess renin secretion can produce secondary aldosteronism, as occurs with renal vascular disease, aortic coarctation, renal parenchymal disease associated with inflammation or scarring or renin-secreting tumors. The first three of these four conditions are common causes of pediatric hypertension, with renal parenchymal disease, renovascular disease and aortic coarctation accounting, respectively, for 60–70%, 5–10% and 10–20% of secondary hypertension in childhood.[4] The simultaneous occurrence of renal artery stenosis and a renal mass has been described, with the mass usually localized within the renal hilum or, less commonly, within the renal parenchyma.[5] Imaging modalities like Doppler ultrasound, computed tomography or magnetic resonance imaging then become necessary for ascertainment and relationship of renal artery stenosis and renal mass. Evaluation of hormones, including plasma renin activity, serum aldosterone concentration and serum cortisol concentration, is a critical component of the evaluation of patients with hypertension and hypokalemic metabolic alkalosis.[6] Excessive production of renin has been observed in several types of renal tumors in children and adults, including juxtaglomerular cell tumors ("reninomas"), Wilms' tumor and rhabdoid tumor of the kidney. Of these, the juxtaglomerular cell tumor is most likely to present with severe hypertension as a primary feature.[7] The first case of reninoma causing hypertension was described in 1967 by Robertson *et al*,[8] juxtaglomerular cell tumour is a rare tumor with an unknown incidence and, till date, 58 cases have

been reported. The tumor appears to have a slight female predominance of 2:1. Such tumors have been diagnosed in patients ranging in age from 6 to 69 years, with an average age at diagnosis of 23.2 years. Appropriate detection and diagnosis of this tumor is imperative because it is a surgically curable cause of severe hypertension, which can be associated with major complications including intestinal ischemia, cerebrovascular accidents, heart disease and death.[9]

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