

CASE REPORTS

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Polyarteritis Nodosa with Renal Artery Stenosis

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Renal angiographic signs in Polyarteritis Nodosa (PAN) vary from aneurysms of medium and small vessels, perfusion defects and delayed emptying of renal arteries. These vascular changes are usually responsible for the hypertension. In this case study stenosis of a main renal artery, an unusual finding in classical PAN, is believed to be the cause of hypertension. Hence renal angiography is essential to define the renal vascular changes and confirm the cause of hypertension.

Key words: Polyarteritis nodosa, Renal artery stenosis, Hypertension.

Polyarteritis nodosa (PAN) is a rare multisystem disease of childhood characterized by necrotising vasculitis

affecting medium sized arteries(1). We present a case of PAN with an unusual association *i.e.*, stenosis of a main renal artery.

Case Report

A three-year-old boy, first born to non-consanguineous parents, previously in good health, developed fever which was insidious in onset, low grade, intermittent, for a period of 3 months. The fever was associated with generalized muscle pain and painful swelling of knees and ankles on both sides, severe enough to prevent the boy from walking. This was followed by blackish discoloration of the skin overlying the same joints as well as the tip

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of fingers in both the hands. He had malaise and significant weight loss. There was no history of hematuria or oliguria.

On examination, the boy was conscious and afebrile. Dry gangrene involving the distal ends of the fourth and fifth finger on the left and the third finger on the right was noted. The skin overlying the anterior aspect of both the knees showed scarring and punched out ulcers. Multiple fine violaceous nodules were noted in both the palms. All the peripheral pulses were well felt and the BP was 120/70 mmHg (<95th percentile). There was no renal bruit.

Investigations revealed the following: Hb 12 g/dL, TC 12,900 cells/cu.mm, DC N 63%, L 37%, ESR 110 mm in one hr and platelet count 6 lakhs/cumm. Serum HBsAg, antineutrophil cytoplasmic antibody (ANCA) and antistreptolysin O (ASLO) titer were negative. Liver enzymes were within normal limits. Urine analysis and renal function tests were normal. Coagulation profile including antiphospholipid antibody did not reveal any abnormality. Ultrasonography of the abdomen and echocardiogram were normal.

In view of the fever, severe myalgia, weight loss, the characteristic gangrenous changes and modular lesions with no evidence of glomerulonephritis the boy was diagnosed to have classical PAN. He was treated with oral prednisolone (2 mg/kg/day) for 8 weeks. As he did not respond to steroids, IV cyclophosphamide (500 mg/m²) was given once in 4 weeks for 6 months in addition to oral prednisolone. Thereafter he was advised oral prednisolone (0.5 mg/kg on alternate day) for 2 years. In addition, acetyl salicylic acid (5 mg/kg/day) was prescribed in view of thrombocytosis and prevention of further thromboembolic complications.

However two years later, the boy presented

again with polyarthritis, fever, myalgia, abdominal pain, malena and gangrenous change of the skin overlying the right ankle. History revealed that he had stopped all drugs 6 months prior to admission. On examination, his blood pressure was found to be high (140/100 mmHg) (>95th percentile) with serial measurements. Investigations revealed the following: Hb 12.5 g/dL, TC 24,000 cells/cu.mm, DC N 70%, L 27%, E 3%, ESR 90 mm in one hr and platelet count 7.4 lakhs/cu.mm. Bleeding time, clotting time, prothrombin time and partial thromboplastin time were within normal limits. Renal function tests were also normal. A flush abdominal aortogram and bilateral selective renal angiography was done which revealed moderate stenosis of the left main renal artery (*Fig.1*). However, there were no aneurysms or segmental narrowing of the intra renal vasculature. In view of the severe abdominal pain and malena, IV methylprednisolone (30 mg/kg/day) was given for 7 days. The hypertension was controlled with enalapril (0.25 mg/kg/day). At discharge, he was relieved of his symptoms and had a BP of 120/80 mmHg. However, the child was subsequently lost for follow up.

Discussion

The initial diagnosis of PAN is clinical. Biopsy of the involved tissue like skin, muscle, sural nerve or kidney may support the diagnosis. Angiography findings include aneurysms, segmental narrowing and variations in the caliber of arteries, together with pruning of peripheral vascular tree often found in the kidney and or liver(2,3). According to Brogan, *et al.*(4) the most reliable non aneurysmal signs in angiography are perfusion defects, presence of collateral arteries, lack of crossing of small peripheral arteries and delayed emptying of small renal arteries. Both PAN and Takayasu arteritis may involve the renal arteries and present with



Fig. 1. Angiography showing stenosis of left renal artery.

severe hypertension due to renal artery stenosis(5). However, arteritis limited to the main renal artery with no involvement of intrarenal vasculature as found in our case is an unusual variant of PAN. This renal artery stenosis may be due to the arteritis(6) or just a coexistent finding and probably contributing to the hypertension. The very young age of onset of PAN in this boy also makes the case interesting. Progressive renal insufficiency

can occur during the acute course of classical PAN due to renal vascular involvement without glomerulonephritis(7). Hence, in a case of PAN with severe hypertension, the exact cause for the hypertension should be ascertained and not presumed to be caused by intrarenal aneurysms.

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