The Turkish Journal of Pediatrics
Volume 17 / Number 1-2 / January - April 1975 / pp. 57 – 64

Gastrointestinal Symptoms in Polyarteritis Nodosa

A Case Report*

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Polyarteritis nodosa (PAN) is a rather rare disease in childhood. It is an arteritis of medium-sized and small arteries. Vascular lesions consist of fibronoid necrosis of the media and infiltration by leucocytes, especially the eosinophil. Since the lesions produced by this disorder may be widely disseminated, it is not surprising that the clinical features are variable, depending on the system involved. Reports describing gastrointestinal complications occurring in this disease have been frequently reported in literature.[1-5]

The purpose of this paper is to present a 6-year-old boy who showed gastrointestinal system complications of PAN and to draw attention to the life-threating intraabdominal complications which may often occur in the absence of gross or easily detectable signs.

Case Report

A six-year-old boy was hospitalized with the complaints of fever, vomiting, unconsciousness and loss of vision. Four months prior to admission he had developed swelling of the left testis. A week later, he was unable to walk because of the swelling of the toes of the right foot. Discoloration of the skin was not associated with this swelling. These complaints subsided within 3-4 days. A month after these first symptoms, the patient complained of abdominal pain, the intensity of which was aggra-

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vated after meals, but there was no vomiting and the pain was not localized in any specific area. 15 days later, the abdominal pain disappeared. A month prior to admission he started to show generalized convulsive seizures, approximately 8-10 times, and left-side hemiplegia developed. Because of this complaint the patient was admitted to another hospital with the diagnosis of meningitis. But laboratory examinations were found to be normal in this aspect. The left-side hemiparesis improved but as the patient could not talk comprehensibly he was referred to our hospital.

Physical examination revealed that his temperature was 37°C, pulse rate 100 per minute; respiration, 16 per minute; and blood pressure 160/130 mmHg. The femoral arterial pulse was bilaterally present. His height was 112 cm and weight 16.5 kg. He was conscious but drowsy and revealed decreased muscular mass. The vision and optic fundi were normal. Nodules, 0.5x0.5 cm in size, and varicose veins were palpated under the skin of the chest and abdomen. A grade 2/VI° systolic murmur was heard along the left sternal border. The liver was 4 cm palpable below the right costal margin. There was neck stiffness and deep tendon reflexes were diminished.

Laboratory Findings: Hemoglobin level was 10.5 gm/dl. The white cell count was 17,800 per cubic mm with 60 percent neutrophils and 40 percent lymphocytes. The platelets were normal. Sedimentation rate was 40 mm per hour. The urinalysis revealed protein and urinary sediment containing 7-8 white cells and sparse red cells. Specific gravity was 1000. Occult blood was detected in the stool in the first examination, but not thereafter.

Blood electrolytes determination showed; Sodium to be 118 mEq/l; potassium 2.1 mEq/l; chloride 101.8 mEq/l and CO_2 content 27.84 mEq/l; BUN was 18 mg/dl.: creatinine 0.4 mg/dl and creatinine clearance 30 cc min/m.^{2}

The SGOT was 60 Units, SGPT 130 units, total bilirubin 12 mg/dl (The direct bilirubin 0.6 mg/dl), thymol 9.1 U., CCF +++ and total protein 6 gm/dl (the albumin 4 gm/dl). The total lipid was 700 mg/dl.

Serum electrophoresis revealed that albumin was 36.2 %; alfa-1 globulin 12.3 %; alfa-2 globulin 25.7 %; beta globulin 4.6 % and gama globulin 21 %. The Coombs test was negative. Antistreptolisin 0 titer was 333 Todd units. CRP, latex, FANA and LE cell were negative.

The electrocardiogram demonstrated nonspecific T wave changes. The X-rays of the chest, abdomen and intravenous pyelogram were all normal. The lumbar puncture was within normal limits.

Since the available muscle biopsy did not contain any medium-sized artery, pathological verification of a collagen disease could not be made. Repeated blood sodium determinations consistently revealed low values despite several corrective attempts. Since the value of the urinary sodium was 59 mEq and potassium 12 mEq per liter, a total 9 gm salt and 2x3 gm KCL was given orally to compensate the urinary electrolytes loss.

Because of the high blood pressure the patient was placed on Hydralazine (1 mg/kg) and Methyldopa (10 mg/kg). Despite this therapy the hypertension was not succesfully controlled. In addition to this antihypertensive treatment, reserpine I.M. (0.07 mg/kg) was added to the therapy which had a partial effect on the high blood pressure.

On the 8th day of his hospitalization, he complained of abdominal pain and vomited once. The next day the patient collapsed abruptly. There was diffuse tenderness and distention of the abdomen. Nasogastric decompression was performed. Blood sodium was 124 mEq and potassium 2.2 mEq per liter and intravenous fluid therapy was started. Hemoglobin level was 7.05 gm/dl and white blood cells were 30,800 per cubic mm. A film of the abdomen disclosed evidence of free air under the diaphragm bilaterally and several air-fluid levels were detected. The central venous pressure was 2 cm of water.

Because of the above-mentioned clinical and laboratory findings, intestinal perforation and peritonitis was diagnosed. Although he was scheduled for emergency surgery, his general condition suddenly deteriorated and he died before any surgical attempt was made. Partial autopsy was done.

Necropsy Findings: The kidney was of normal size and weighed 80 gm. The subcapsular surface showed evidence of infarction, (Figure 1). Upon cutting the surface, infarcts were also seen. At the corticomedullary junction, arteries appeared very prominent and thrombosed vessels were observed (Figure 2). Swellings and thrombosis were not present on the main divisions of the renal artery and the segmental branches.

Opon microscopical examination some of the medium-sized vessels proved to be involved in arteritis, most of which were healing or healed (Figures 3-4). Associated tubular atrophy was observed. Most of the glomeruli appeared normal or minimally abnormal, with only a slight, irregular increase in mesangial cells and mesangial matrix.

These findings are consistent with the diagnosis of the classic form of polyarteritis nodosa.

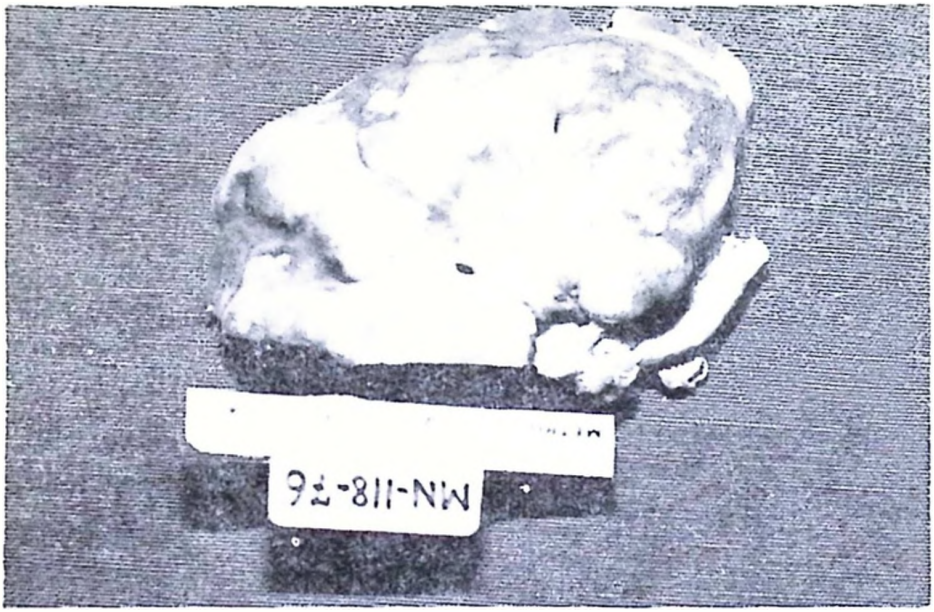

Figure 1

Typically scarred subcapsular surface of kidney in healing polyarthritis. Raised areas consist of healthy hypertrophic parenchyma, while depressed areas show considerable ischemia.

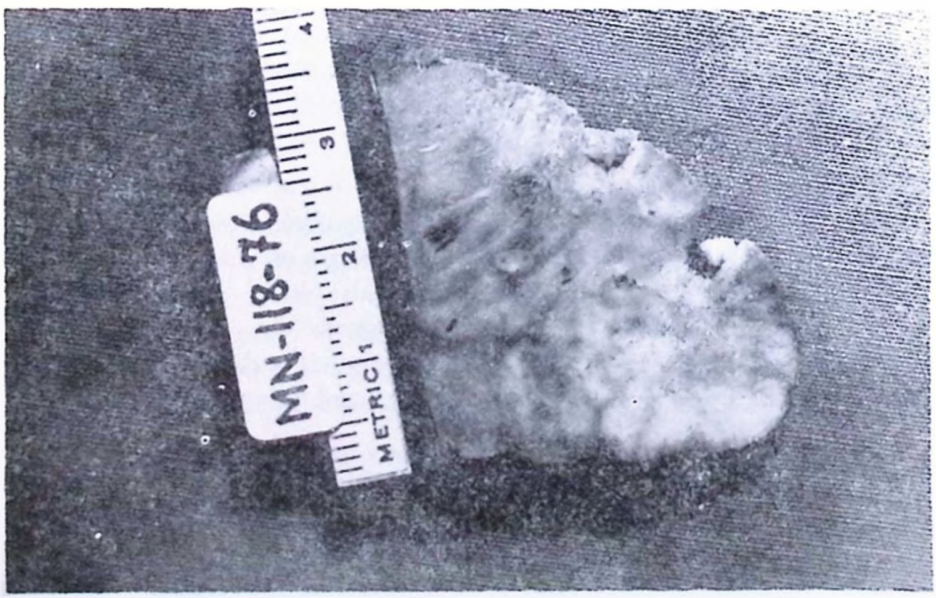

Figure 2

Cut-surface of the kidney showing numerous infarcted areas in the cortex and dark thrombosed arteries at the corticomedullary junction.

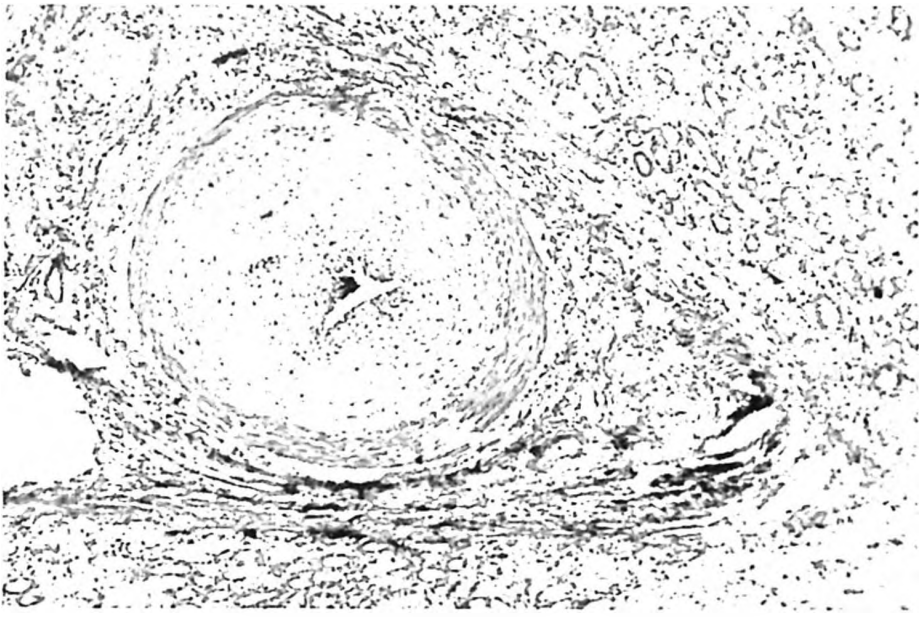

Figure 3

Healing phase in a segment of arcuate artery. The intima is considerably thickened by fibroblastic cells. The lumen is almost completely obliterated. (X 100).

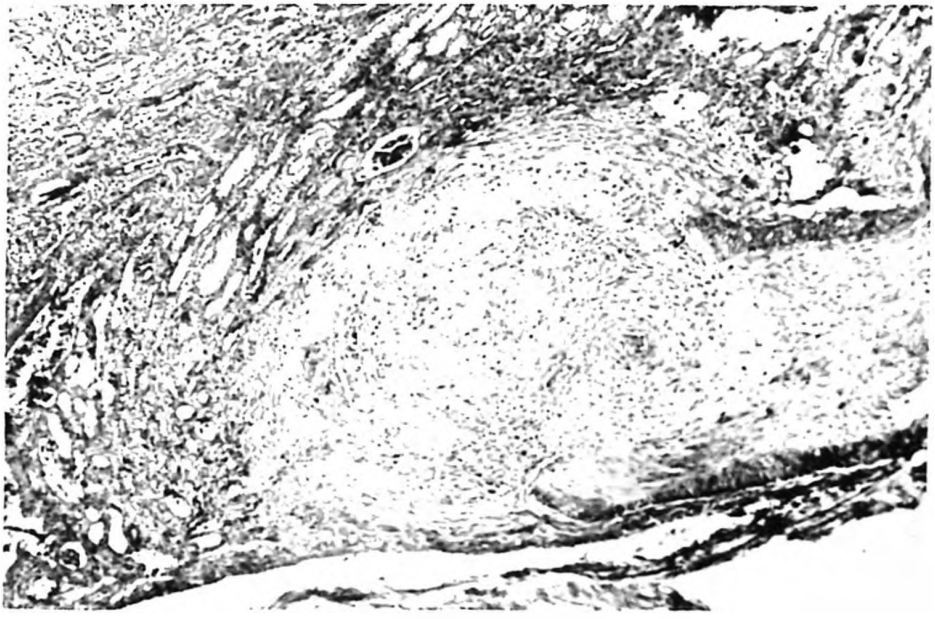

Figure 4

Arcuate artery in chronic healed stage. There is complete fibrous replacement and obliteration of lumen. (X 100).

Discussion

PAN can occur from infancy to old age with a peak incidence in the fifth and sixth decades of life. The male to female ratio has been estimated at from 2 to 3:1.[6] Because of the impaired arterial supply in different organs this disease produces diverse clinical symptoms.

Our patient's clinical manifestations and histological findings were typical of PAN. Systemic illness (such as fever and weakness), cutaneous manifestations, muscle weakness, arthiritis, seizures, paralysis, loss of vision, and orchitis are frequently encountered in polyarteritis.

Arthralgia and myalgia are common in PAN. Arthralgia is migratory, generally without swelling and due to small localized arterial lesions rather than extensive synovitis.

Cutaneous involvement is seen approximately in 25 percent of those affected with PAN 6. These manifestations varied from polymorphic exenthema (purpuric, urticarial and multiform in character) to severe subcutaneous hemorrhage. A most characteristic, but uncommon finding is cutaneous and subcutaneous nodules. The nodules are usually movable, may regress in days or persist for months. Size of the nodules vary from a pea to a walnut and occur at any time in the course of the disease.

Different cardiac manifestations can be seen, but pulmonary involvement is probably infrequent in cases of generalized PAN. Involvement of the ovaries and epididymis is frequent.

Neurologic manifestations are generally late occurrences in the course of PAN. Headache, convulsive seizures, papillitis, retinal hemorrhages and exudates, meningeal irritations, neuritis and peripheral neuropathy can all be seen in the course of PAN.

Arteritis occurs in the kidney with great frequency in different series (70.8 and 82 percent). Glomerulonephritis, which includes glomeruli showing necrotizing or proliferative changes or scarred glomeruli, was found in 18-83 % of patients with PAN.[5,7] These wide discrepancies may be attributed to the varying criteria used to define glomerulonephritis. Membranoproliferative glomerulonephritis, possibly produced by circulating immune complexes, are also described in PAN.[8] In general 25-30 percent of all patients with polyarteritis die from renal failure. When the kidney is affected, the symptoms and signs will vary according to the type of renal involvement. These clinical features vary from merely proteinuria and cells in the urine, to acute oliguric renal fail-

ure. Acute oliguric renal failure associated with this condition is rare, accounting for only 5-10 % of the cases. The outcome of this disease in all these patients was uniformly fatal.[9]

Hypertension is found with great frequency in PAN, and figures in the literature vary between 44-49 percent.[7] Opinions for the pathogenesis of the hypertension in this disease are different. But the theory that the vascular lesions may initiate hypertension is more probable than others.[7]

Our case showed electrolyte imbalance without azotemia, and especially an urinary electrolyte loss. This finding leads us to consider the possibility of tubular disturbance. In PAN these types of clinical and laboratory changes occur in the classic form of PAN rather than in the microscopic form. It was also confirmed by histopathological findings of the kidney, that this case was a classic form of PAN.

In the patients with collagen vascular disease, life-threatening intra-abdominal complications are not uncommon.[1-6] Matolo and Albo conducted a series with a total of 122 patients, of whom 26 (21.3 %) had major gastrointestinal system complications.[1] Of the patients in this series with SLE, 27.5% showed gastrointestinal system complications; with PAN 36.0 %; with scleroderma 10.7 % and from those with dermatomyositis there were none. In PAN the inflammatory change in small vessels may lead to thrombosis and ulceration of the gastrointestinal tract mucosa. The following types of complications in PAN have been encountered; gastro-intestinal ulceration with hemorrhage 28.2 %; GI ulceration without hemorrhage 10.3 %; peritonitis 13.5 %; vascular occlusion 10.3 %; acute pancreatitis 6.9 %; colitis 6.9 %; abdominal pain of undetermined etiology 10.3 % and gastrointestinal obstruction 3.4 %. As can be seen, gastrointestinal ulceration with hemorrhage accounted for the majority of the surgical complications (28.2 %).

The majority of these patients were receiving steroid therapy which often masked the severity of their complication. For this reason it is essential to be aware of the GI system complications in collagen vascular disease and to be alert for an early diagnosis; properly timed and selected therapy (medical or surgical) are necessary.

Summary

The case of a 6-year-old boy with gastrointestinal system complications of polyarteritis nodosa is presented and the gastro-intestinal system complications of this disease are discussed.

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