The Turkish Journal of Pediatrics 1996; 38: 101-105

DIAGNOSTIC VALUE OF RENAL ARTERIOGRAPHY IN POLYARTERITIS NODOSA*

*Neslihan Tekin MD***, *Nurdan Kural MD****, *Tamer Kaya MD*****

SUMMARY: Tekin N, Kural N, Kaya T. (Departments of Pediatrics and Radiology, Osmangazi University Faculty of Medicine, Eskişehir, Turkey). Diagnostic value of renal arteriography in polyarteritis nodosa. Turk J Pediatr 1986; 38: 101-105.

Polyarteritis Nodosa (PAN) is a rare disease in childhood. No single pattern of clinical presentation characterizes this disease, but abdominal pain, central or peripheral nervous system disease, arthritis, myalgia and skin lesions occur at some time during the course of the illness. In this case a 16-year-old boy who presented with abdominal pain, elevated sedimentation rate associated with hypertension, and a high level of renin, all of which were detected during his hospitalization, suggested the diagnosis of PAN, and renal angiography was performed. Characteristic renal aneurysms were visualized and the diagnosis was confirmed. *Key words: renal arteriography, renal aneurysms, polyarteritis nodosa, hypertension.*

Polyarteritis nodosa (PAN) is a well-known form of necrotizing angiitis which is characterized by focal necrosis of the walls of small- and medium-sized arteries[1-3]. The angiitis of polyarteritis affects various organs singly or in combination. Kidney, skin, joints, heart, peripheral nerves and the gastrointestinal tract are the most common sites of involvement[4, 5]. Lung, skeletal muscle and central nervous system involvement can occur at any time during the course of the disease.

The diagnosis of PAN is based on clinical evidence of multi-organ involvement corroborated by demonstration of vasculitis by either histopathologic changes on biopsies of skin, muscle or kidney, electrocardiography, electromyography or arteriography[6]. It is a rare disease of childhood and in most cases diagnosis is not confirmed early in the course of the disease because PAN closely mimics many diseases, especially the other collagen tissue disorders. In some cases biopsy is not diagnostic.

In this case the diagnosis of PAN was established by demonstrating aneurysms in the renal arteriography of a 16-year-old patient. We report this case to draw attention to the diagnostic value of renal arteriography in PAN-suspected cases, especially when, histopathological evaluation is not helpful in identifying the diagnosis.

* From the Departments of Pediatrics and Radiology, Osmangazi University Faculty of Medicine, Eskişehir.

** Assistant Professor of Pediatrics, Osmangazi University Faculty of Medicine.

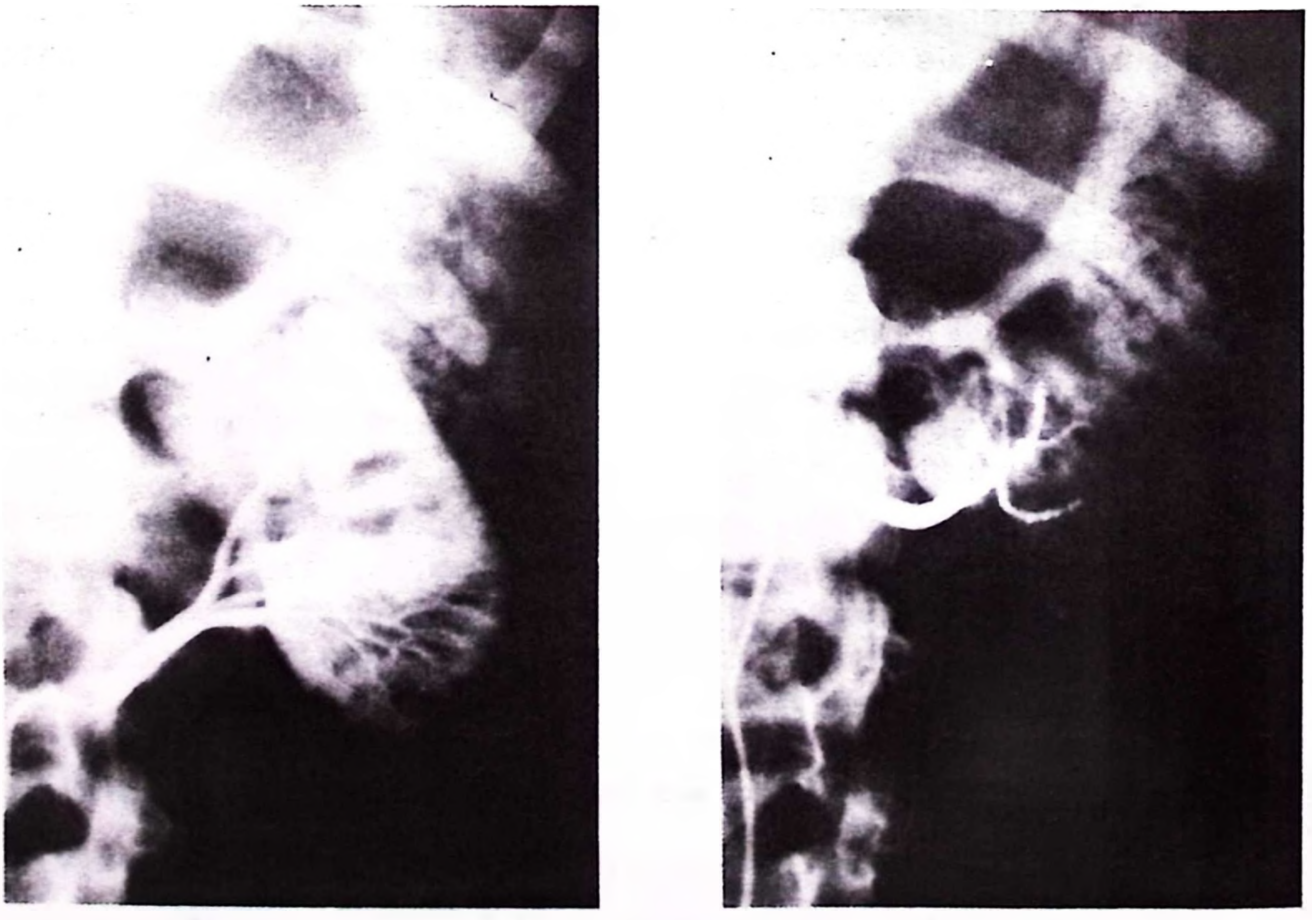
*** Professor of Pediatrics, Osmangazi University Faculty of Medicine.

**** Associate Professor of Radiology, Osmangazi University Faculty of Medicine.

Case Report

A 16-year-old boy presented with abdominal pain. He had been suffering continuous pain around his umbilicus which extended towards both costolumbar regions for 14 days.

Vital signs: blood pressure 120/80 mmHg, heart rate 64/min, temperature 36.7 °C. Tenderness was present during palpation of both of the costolumbar regions. His physical examination was otherwise unremarkable. The hemogram revealed Hb 13.2 g/dl, platelets 355.000/mm^3, total white cell count 12.100/mm^3 with 75% neutrophils and 25% lymphocytes. Erythrocyte sedimentation rate 65 mm/h, Fibrinogen 575 mg/dl. Urinalysis repeated five times was normal. BUN 20 mg/dl, Cr 1 mg/dl, blood glucose 101 mg/dl, Ca 10 mg/dl, Na 141 mEq/L, K 4.8 mEq/L, SGOT 30 U/L (7-39), SGPT 25 U/L (2-54), Alkaline phosphatase 65 U/L (41-133), total protein 7.5 g/dl, albumin 3.9 g/dl, amylase 120 mg/dl (50-180), renin 7 ng/ml (1.5-5.7), aldosterone 52 ng/dl (12-125), ASO 833 Todd U, CRP 24 mg/dl, HBsAg (-), IgG 2400 mg/dl (800-1700), IgA 266 mg/dl (85-490), IgM 99.3 (50-370) mg/dl, C3 89.8 mg/dl (50-90), C4 46.1 mg/dl (10-40), ANA (-), anti DNA 4.26 IU/ml (0-6). No pathogenic microorganism was detected in cultures taken from the throat, urine



Figs: 1 and 2: Renal arteriography showing the characteristic aneurysms.

or stool. Chest roentgenogram, ultrasonography and computed tomography of the abdomen, biopsy of the skin, subcutaneous tissue and muscle were all normal.

During the course of hospitalization hypertension was detected up to 160/110 mmHg and persisted at high levels. When a high level of renin was found, renal artery angiography was performed to evaluate the case. Renal aneurysms characteristic of PAN were visualized (Figs. 1 and 2). Steroid therapy was administered and the patient was followed up.

Discussion

The diagnosis of polyarteritis nodosa is based on clinical evidence of multisystemic involvement, biopsies of the kidney, muscle or skin and selective arteriography of the kidney, hepatic or mesenteric arteries[4]. As it is known that PAN in children is such a serious and often rapidly fatal disease, early recognition and treatment are important. Diagnosis with only clinical features may not be correct. If clinical or laboratory evidence suggests the diagnosis of PAN, biopsy or arteriography of selected tissues should be performed.

Özen et al.[7] established two major and ten minor findings of the disease as diagnostic criteria of PAN in children. The presence of five of these 12 criteria, including at least one major criterion, has been used to indicate the diagnosis of PAN, but in most of the series the diagnosis needed to be confirmed by either biopsy or arteriography.

Abdominal pain, the most common gastrointestinal symptom in PAN, was our patient's only complaint. In Pediatric PAN series, it has been reported that abdominal pain was a complaint in 20 of 25 in one group and 10 of 11 in the other[5, 8]. When the other series are combined, abdominal pain was present in 67 percent of patients[5].

Renal involvement in PAN can conveniently be divided into two distinct types: one affecting medium-sized extraglomerular vessels (classical macroscopic PAN) and the other characterized by microscopic arteriolar/capillary involvement and glomerulonephritis[9]. Aneurysmal dilatation is classically confined to medium-sized vessels and is seen in angiography. Although hypertension occurs more frequently among those individuals whose arterial lesions involve medium-size vessels, it may also develop as a result of glomerular involvement[9]. In this case because the urinary findings and renal function studies were normal, we believed that the hypertension originated from the involvement of medium-sized extraglomerular vessels. That was proved by the elevated renin level and formation of renal artery aneurysms. The incidence of hypertension varies among the series. All 11 children with PAN reported by Blau et al.[8] had hypertension. In the report of Özen et al.[7], hypertension developed in 20 out of 31 cases. In a study of 36 PAN cases, hypertension was the second most common clinical with an incidence of 83 percent[5].

Elevated ESR, CRP and mild leukocytosis were present in our case, which may be accepted as nonspecific laboratory findings of PAN. However, the features described here are not enough to confirm the diagnosis. The demonstration of perinuclear-anti-neutrophil cytoplasmic antibodies (p-ANCA), which is usually associated with PAN and other vasculitides, would be more helpful in making a specific diagnosis[9].

Typical involvement of medium-sized muscular arteries and small arteries could not be demonstrated in our patient's skin biopsy. Biopsies of skin or muscle are the easiest to obtain but are frequently normal, especially if the specimen is taken from an uninvolved site. Because of normal urinary findings and renal functions we did not consider doing a kidney biopsy. Renal arteriography was performed, and characteristic renal aneurysms were visualized. The first case with PAN diagnosed by renal angiography in the pediatric age-group was reported in 1972 by McLain et al.[10] but when compared with our case he had several other manifestations of the disease, even myocardial infarction. Angiography has a 90, 94 percent sensitivity in adult patients with PAN[11]. The abnormalities include both arterial aneurysms and occlusions. The mechanism of formation of aneurysms is the weakened wall which tends to bulge because of fibrinoid necrosis, and inflammation with polymorphonuclear leukocytes, eosinophils, and round cells[12].

In this case we draw attention to the diagnostic value of arteriography when the clinical features are not clear and biopsy does not show the characteristic histopathological changes.

REFERENCES

1. Ettlinger RE, Nelson AM, Burke EC, Lie JT. Polyarteritis nodosa in childhood. Arhtritis Rheum 1979; 22: 820-825.
2. Guillevin L, Le Thi Huong Du, Godeau P, Jais P, Wechsler B. Clinical findings and prognosis of polyarteritis nodosa and Churg-Strauss angiitis: a study in 165 patients. Br J Rheum 1988; 27: 258-264.
3. Fink CW. Vasculitis. Pediatr Clin North Am 1986; 33: 1203-1219.
4. Reimold EW, Weinberg AG, Fink CW, Battles ND. Polyarteritis in children. Am J Dis Child 1976; 130: 534-541.
5. Cassidy JT, Petty RE. Vasculitis. In: Cassidy JT, Petty RE (eds). Textbook of Pediatric Rheumotolgy (2nd ed). New York: Churchill Livingstone; 1990: 377-424.
6. Magilavy DB, Petty RE, Cassidy JT, Sullivann DB. A syndrome of childhood polyarteritis. J Pediatr 1977; 91: 25-30.
7. Özen S, Beşbaş N. Saatçi U, Bakkaloğlu A. Diagnostic criteria for polyarteritis nodosa in childhood. J Pediatr 1992; 120: 206-209.
8. Blau EB, Morris RF, Yunis EJ. Polyarteritis nodosa in older children. Pediatrics 1977; 60: 227-234.
9. Roberti I, Reisman L, Churg J. Vasculitis in childhood. Pediatr Nephrol 1993; 7: 479-489.

10. McLain LG, Bookstein JJ, Kelsch RC. Polyarteritis nodosa diagnostic by renal arteriography. J Pediatr 1972; 80: 1032-1034.

11. Lightfoot RW Jr, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of polyarteritis nodosa. Arthritis Rheum 1990; 33: 1088-1093.

12. Schaller JG. Polyarteritis Nodosa. In: Behrman RE, Kliegman RM, Nelson WE, Vaughan VC. III (eds). Nelson Textbook of Pediatrics (14th ed). Philadelphia: W.B. Saunders Co; 1992: 631.