

CUTANEOUS POLYARTERITIS NODOSA: A REPORT OF TWO CASES

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Abstract - Polyarteritis nodosa is a multisystem necrotizing vasculitis of small and medium size muscular arteries in which involvement of the renal and visceral arteries is characteristic. The etiology is unknown but hepatitis B antigenemia is found in 30% of patients. Drug exposures and hypersensitivity reactions are other possible causes. Excellent therapeutic results have been reported in classic polyarteritis nodosa with the combination of corticosteroids and cytotoxic agents. In this article, 2 cases of cutaneous polyarteritis nodosa are reported
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Key words: Cutaneous Polyarteritis Nodosa, artery, vessel walls, inflammation, corticosteroid

INTRODUCTION

Polyarteritis in its classic form was first delineated in 1866 by Kussmaul and Maier and became known as Kussmaul-Maier disease. Over the years the full spectrum of the clinical and pathological manifestations of the disease were recorded by various investigators and new names were coined for it. Although the etiology of the disease is still not quite clear, there is enough evidence to prove that immunologic mechanisms play a considerable role in its pathogenesis and new horizons are opened for the study and management of this disorder. The wide range of the clinical manifestations and pathological findings in the disease, results from multi system necrotizing vasculitis, and presents many diagnostic and therapeutic challenges to the clinician. Although the disease is usually systemic with multi - organ involvement, it is not uncommon to encounter patients at early stages of the disease or with only a single organ involvement. Since early diagnosis and proper management can drastically improve the overall prognosis and diminish morbidity and mortality, it is critical for the clinician to be on alert for the not- so- typical presentations of this illness

(1,2).

CASE 1

AJ. was a six years old boy admitted because of diffuse painful red - violet skin lesions four months of duration. Significant in his past history, were episodes of urticaria and vesicular skin lesions which improved by medical treatment. His parents, brother and two sisters were reportedly in good health. On exam multiple red - violet skin lesions of different size were noted under the eyelids, over the nose, on both sides of the trunk and thighs, on the left hypothenar eminence and on the first right finger; the lesions were firm tender nodules with the old ones being yellow and the majority red-violet in colour. The clinical examination was otherwise completely normal. Blood pressure (BP): 100/50 mmHg. weight (W): 16.7 kg, white blood cell count (WBC): 8600, Seg: 60%, Ly: 30%, Mono: 5%, Eo: 5%, hemoglobin (Hb): 12.6, hematocrit (Hct): 42%, Platelet: normal ESR: 30 - 70 mm. BUN: 30 mg / dL, serum total protein: 6.7 g / dl with electrophoresis showing an increase in gamma globulin and alpha 2 globulin. Anti - streptolysin O (ASO): 625 Todd units, hepatitis B surface antigen (HBs Ag), lupus erythematosus (LE) cell, latex and PPD were all negative. Chest X ray, ECG, urinalysis, stool exam and evaluation for anti - nuclear antibodies (ANA) and complement measurement were all normal.

A biopsy from one of the skin lesions revealed normal epiderm with remarkable pathologic changes in the small arteries of the subdermal layer in the form of scar and fibrosis of the media and adventitia which was diagnostic of polyarteritis.

The patient was treated with prednisolone 2 mg/kg/day for two weeks in the hospital and six weeks at home. Then he returned to the hospital with herpes zoster and prednisolone was discontinued. One month

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later, and the old skin lesions had healed without any new ones. He was placed on prednisolone again. The next examination performed after 3 months revealed complete recovery without any skin lesion lesions and ESR fell to 5 mm.

CASE 2

P. M. was a seven year old girl admitted because of arthralgia of the right elbow associated with fever and weight loss of three month duration. Her illness first started with an irregular fever which did not respond to antibiotic therapy. The parents were not relatives. They had eight children, two of whom had died from unknown causes. Hb 5.8 g /dl, Hct: 23%, WBC 38000, Neut: 80%, Eo: 1%, ESR ranged from 100 to 145 mm in multiple examinations. Serum protein electrophoresis revealed albumin of 26.2%, α_2 globulin of 10.8% and γ globulins of 43.5%. LE preparation, HBs Ag, PPD, blood cultures, Wright and widal tests were all negative. Urinalysis showed mild proteinuria. ANA and complement measurements were normal. X Rays of the chest, arm, and spine did not show any abnormality. A few days after admission firm nodules appeared over the skin of her right hand and arm and both wrists became painful. Biopsy from one of the nodules showed leukocyte infiltration of the arterial wall with edematous changes of the adventitia. These changes were interpreted diagnostic for polyarteritis. The patient was treated with corticosteroids.

DISCUSSION

Polyarteritis is a relatively uncommon disease. It is more common in men than in women. The mean age of onset is 45 years; male to female ratio is about 2.5 to 1. There is no known racial or familial predisposition.

Polyarteritis in children has at least three variants. The first, a generalized disease, is seen both in children and adults. In the second, infantile polyarteritis nodosa, coronary artery involvement is common, and infants present with congestive heart failure. The third, mucocutaneous lymph node syndrome, appears similar to infantile polyarteritis nodosa but occurs in older

children and has distinct mucosal and cutaneous lesions. The disease is very rare in childhood. In older children it resembles the adult form of the disorder but a distinctive syndrome is seen in infantile polyarteritis. The latter is quite rare and occurs in patients younger than 12 months of age(3). Interested reader is referred to the reports of 10 cases in children under age 10, and 14 cases in patients during childhood from Japan (4 - 5). The exact etiology of the disease is still unknown but a hypersensitivity reaction is a probable cause. The presence of hepatitis B antigenemia in approximately 30% of the patients with systemic vasculitis, particularly of the classic PAN type, together with the isolation of circulating immune- complexes composed of HB antigen and IgM as well as the demonstration by immuno fluorescence exposure of HB antigen, IgM and complement in the blood vessel walls strongly suggest the role of an immunologic phenomenon in the pathogenesis of this disease. The disease has also been reported following to certain drugs. (6). There is some relation between rheumatoid arthritis and PAN and rheumatoid factor is positive in some cases. Some investigators believe that there is a relation between PAN and acute rheumatic fever and Aschoff nodules are seen in a few cases of the former disease (7, 8).

The necrotizing inflammation involves medium and small arteries, adjacent veins and occasionally arterioles and venules but not capillaries. Microscopically the changes in the vessels may be divided into acute, healing, and healed stages. Within the same case of the classic type of polyarteritis one frequently sees all three stages although in other cases one stage may predominate. The acute lesions are characterized by fibrinoid necrosis which may affect only the intima, but often extends to involve the full thickness of the arterial wall particularly in small arteries. Healing lesions are characterized by fibroblastic proliferation in addition to the continuing necrotizing process. The healed lesions consist merely of marked fibrotic thickening of the affected arterial wall. Although these stages have been described separately all three may coexist in different foci either within the same vessel or in different vessels (9).

Clinical presentation is different in patients,

because different organ involvement, produces different signs and symptoms. The initial manifestations are fever, weight loss, tachycardia and pain. Focal manifestation include GI, skin, joints and peripheral nerve involvement. Occasionally the patient presents with fever, leukocytosis, abdominal pain which can be mistaken with acute abdomen. Three fourth of the patients have kidney involvement. The renal manifestations usually present as acute glomerulonephritis. The patient may also present with hypertension, myocardial infarction and pulmonary infiltration. In children the urinalysis may show proteinuria and pyuria which are resistant to treatment. Periarteritis of the renal arteries as well as glomerulitis are responsible for hypertension which is an ominous sign, seen in 60% of cases. Abdominal pain is very common and associated complaints such as nausea, vomiting and diarrhea may also be present.

Mesentric thrombosis may lead to non- infectious peritonitis which can be mistaken with acute abdomen. Involvement of the appendix and pancreas arteries are not uncommon and can mimic clinical manifestation of appendicitis or pancreatitis. One half of the cases of hepatic necrosis and jaundice may be due to periarteritis nodosa. Myocarditis is the common cardiac manifestation of the disease. In children, heart failure and pericarditis are rare, although involvement of coronary arteries and myocardial infarction are not uncommon. Nervous system involvement is seen in about 30-60% of the cases mainly affecting the peripheral and central nerves. Arthralgia is seen mainly in large and moderate size joints and can mimic acute rheumatic fever or rheumatoid arthritis. Skin nodules are seen in 25% of cases and more often on the inner aspect of the arm and hand. Occasionally vesicular lesion, urticaria and angiomatous edema of the skin, and rarely gangrene of the fingers have been reported. Anemia is usually due to hemorrhage and ESR is almost always elevated. Eye involvement, in the form of nodules under conjunctiva, retinal hemorrhage and exudates are reported.

Prognosis of the illness is variable. Death usually results from renal failure, GI complications, particularly bowel infarcts and perforation, and from cardiac causes

(10, 11).

The 5 year survival rate of untreated patients has been reported to be 13 percent. Corticosteroid therapy alone may increase this figure to over 40% and extremely favorable results have been reported in classic PAN with the combination of prednisolone 1 mg/kg/day and cyclophosphamide 2 mg/kg/day. This regimen has been reported to result in long-term remission in up to 90% of cases, even following discontinuation of therapy. Isolated reports have indicated favorable responses in PAN using plasmapheresis together with corticosteroids and cytotoxic agents (2-10).

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