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Case Report

Successful Treatment of Spontaneous Splenic Rupture in a Patient with Polyarteritis Nodosa

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Abstract

Polyarteritis nodosa (PAN) is a rare necrotizing vasculitis presenting with multiple organ manifestations. We herein describe a case presented with splenic subcapsular hematoma, after nontraumatic splenic rupture and successfully treated with a combination of pulse steroids and cyclophosphamide therapy.

Keywords: Polyarteritis nodosa; Splenic rupture; Gastrointestinal

1. Introduction

Polyarteritis nodosa (PAN) is a rare syndrome, predominantly affects medium sized arteries in various organs [1]. The skin and peripheral nervous system are the most frequently involved target organs followed by the gastrointestinal tract and the kidneys which indicate a poor prognosis [1]. Our case presented with pure cutaneous manifestations seven years ago, systemic involvement was excluded by MR angiography in that period. But now he presented with gastrointestinal involvement. We emphasized the presentation and treatment of this case.

2. Case Report

A 43 year old man, with obstructive cerebrovascular disease history, was diagnosed cutaneous PAN with a biopsy on erythema nodosum-like skin lesions seven years ago. Pathological findings were fibrinoid necrotic vasculitis of middle and small arteries. The patient was followed in remission, treatment with steroids and methotrexate. He presents with sudden onset, severe, diffuse abdominal pain of 18 hours' duration and evaluated in the emergency room, diagnosed with spontaneous splenic rupture and acute abdomen. Abdominal examination revealed severe tenderness in

the upper umbilical region with rebound tenderness. Diagnostic imaging with computed tomography showed 1 cm liver hemangioma, hepatic artery aneurism and perisplenic high-density fluid in hemorrhagic characteristics. Investigations revealed elevated C reactive protein, erythrocyte sedimentation rate, neutrophilic leukocytosis. Considering the main organ involvement, treatment with both pulse steroids (methylprednisolone 1000 mg/day, three days followed by 64 mg/day) and cyclophosphamide (1000 mg/month six times) was administered, leading to significant clinical improvement. In the first month of the treatment, we observed that the hematoma was organized in images and the patient is stable. After six cycles of cyclophosphamide treatment, six months after disease onset; PAN was inactive. Long term maintenance therapy of the patient continued with oral azathioprine. Today the patient is in good clinical condition with no abdominal pain, no fatigue, no skin rash. Acute phase reactants returned to the normal range.

3. Discussion

Patients diagnosed polyarteritis nodosa (PAN), may present with cutaneous and life threatening multiple organ manifestations such as gastrointestinal, cardiac, neurologic system involvement [2]. Histopathology showed the inflammation and fibrinoid necrosis in the entire arterial wall, which is a typical feature of PAN. Also deposition of polymorphonuclear leucocytes and antigen-antibody complexes in the arterial wall weakens the vessels, prepares the ground for aneurysm formation and eventual rupture [3]. Gastrointestinal (GI) involvement with ischemia and hemorrhage can cause acute abdomen. Surgery may be necessary due to bowel perforation or massive GİS bleeding [4]. High mortality rates can be seen in patients presenting with acute abdomen despite combined medical and surgical treatment [5]. Limited form of the disease, cutaneous PAN is more frequent than systemic disease, typically have benign clinical course and its etiology is unknown. Progressing of cutaneous form to the systemic PAN is rarely seen [6]. Treatment is dependent on the severity of disease which includes corticosteroids with the addition of immunosuppressive agents. Combination of high dose steroids and cyclophosphamide treatment was recommended for remission induction in patients with moderate to severe systemic vasculitides (any evidence of renal insufficiency, gastrointestinal, cardiac, or neurologic involvement) followed by long term maintenance therapy with steroid sparing drugs such as azathiopurine, mycophenolate mofetil and methotrexate [7].

4. Conclusion

Polyarteritis nodosa (PAN) may present with life threatening organ manifestations. We describe the case of a patient with PAN presented with bleeding due to splenic rupture, successfully treated with a combination of pulse steroids and cyclophosphamide therapy. Failures of response to immunosuppressive medications and radiological intervention can lead to high mortality or permanent organ damage. Timely diagnosis and aggressive treatment are keys to a successful outcome.

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