Polyarteritis nodosa presenting as profuse gastrointestinal bleeding : a case report

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ABSTRACT

We report the case of a five years old boy presented with abundant hematemesis. Following resuscitation, upper gastrointestinal endoscopy was performed and showed a duodenal ulcer. Under medical treatment, the haematemesis recurred with hemorrhagic shock, requiring a laparotomy. Preoperatively, a bleeding ruptured aneurysm of the right renal artery was discovered. The postoperative course was marked by the occurrence of High blood pressure and peripheral neuropathy. Arteriography confirmed aneurismal dilatation of the right renal artery. Erythrocyte sedimentation rate was of 110mm/hour.

Antineutrophil cytoplasmic antibodies, serum cryoglobulins and anti-DNA antibodies were negative. Complement levels were within normal ranges. The diagnosis of PAN was made. The patient was managed by prednisone and cyclophosphamide with good outcome.

In the cases of seemingly obscure gastrointestinal bleeding or the occurrence of duodenal ulcer in children without risk factors for peptic ulcer, the diagnosis of necrotizing vasculitis should be suspected. At least, Erythrocyte sedimentation rate should be early performed in these patients.

Keywords : Vasculitis ; Polyarteritis nodosa ; ANCA ; Childhood

INTRODUCTION :

Polyarteritis nodosa (PAN) is a rare multisystem disease characterized by systemic necrotizing arteritis of small- and medium-sized arteries (1). It may affect many organ systems and thus it can present with a broad array of symptoms. These manifestations result from ischemic damage to affected organs. This condition affects adults more frequently than children and most cases occur between the ages of 40 and 60 years (2). Early onset PAN is extremely rare which makes the diagnosis unrecognized by pediatricians.

We report an atypical case of PAN revealed by digestive hemorrhage in a five year old child.

Case report : A five years-old boy was referred to the hospital with abundant gastrointestinal bleeding associating hematemesis and rectal bleeding. He had no family history of peptic ulcer disease. He was well until two weeks before admission, when he developed abdominal pain. He did not take non-steroidal anti-inflammatory drugs and had no associated complaints.

On examination, he was eutrophic. He had a cutaneous paleness without jaundice. Body temperature was of 37 °C. He had a tachycardia with a heart rate of 140 beats per minutes and blood pressure of 90/60 mmHg. Respiratory rate was of 22/mn. He had a soft and compressible abdomen, without hepatosplenomegaly or collateral venous circulation. Complete blood count showed a regenerative microcytic hypochromic anemia at 9 g / dL. The prothrombin time was 90 % (normal range: 70-100%).

performed and showed a rounded formation with a diameter of about 1 cm and a necrotic edge located in the lumen of the postbulbar zone of the duodenum. Given the existence of significant bleeding, biopsies were not performed. Abdominal CT scan objectified peri-duodenal infiltration suggesting a perforated and stuffy ulcer (figure 1).

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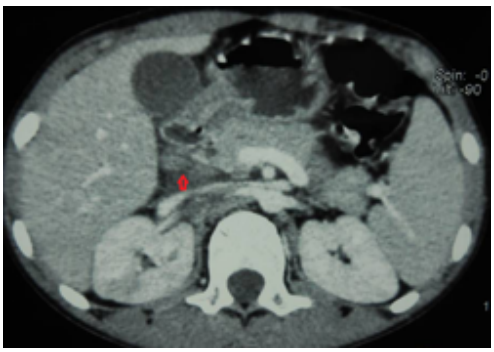

figure 1 : Abdominal CT scan showing peri-duodenal infiltration suggesting a perforated and stuffy ulcer.

Initial management was based on concomitant therapy associating Omeprazole: 2 mg/Kg/day, Clarithromycin: 30 mg/kg/day, Amoxicillin: 60 mg/kg/day, and Nitroimidazole: 30mg/kg/day prescribed for 14 days.
Despite regular medication intake, the gastrointestinal bleeding recurred 12 days later and the patient presented with haemorrhagic shock requiring laparotomy. During surgery, there was a bleeding that was secondary to a ruptured right renal artery aneurysm. The aneurysm was repaired by in situ reconstruction and the perforated ulcer was managed by simple sutures without omental patch.
The postoperative course was marked by hemodynamic stability but the occurrence of High blood pressure at 180/100 mm Hg and paresthesia of the left lower limb on the third post-operative day.
Diuresis, renal function and serum electrolytes were normal. There was no proteinuria or hematuria. Because of the occurrence of severe headache and hypertension, a brain CT scan was performed and showed hyperintensity in cortical, subcortical and left frontal areas evoking posterior reversible encephalopathy syndrome (PRES) (Figure 2).

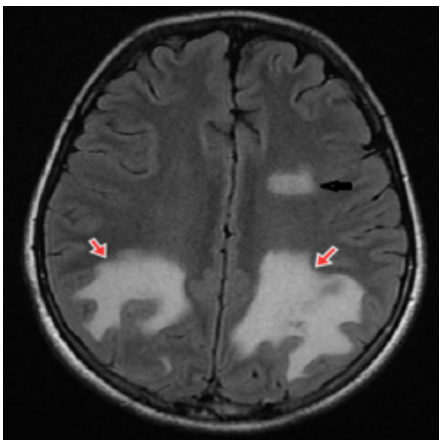

figure 2 : Brain CT scan showing hyperintensity in cortical, subcortical and left frontal areas evoking posterior reversible encephalopathy syndrome.

Electromyogram revealed total degeneration of the external popliteal sciatic nerve and subtotal degeneration of the internal popliteal sciatic fibers. Abdominal CT scan discovered an infraction of the right kidney (Figure 3) and hypodensity of the right gluteal muscles evoking myositis (Figure 4).

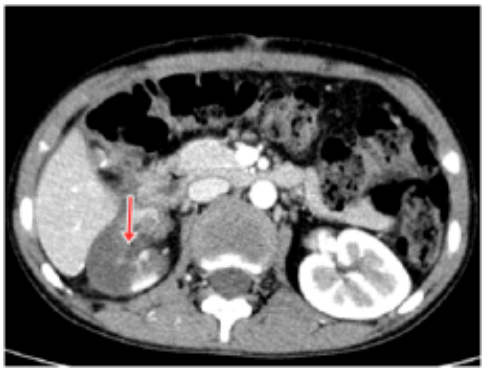

figure 3 : Abdominal CT scan showing an infraction of the right kidney.

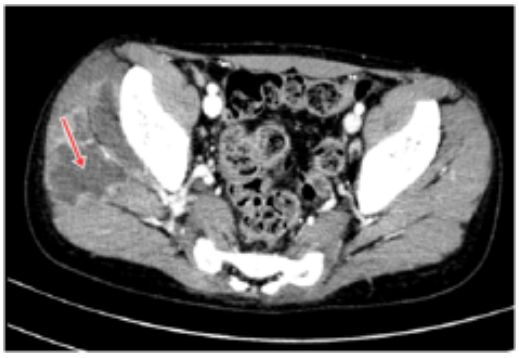

figure 4 : CT scan showing hypodensity of the right gluteal muscles evoking myositis.

To explore renal damage, the patient underwent a selective arteriography which confirmed aneurysmal dilatations of the right renal artery (Figure 5).

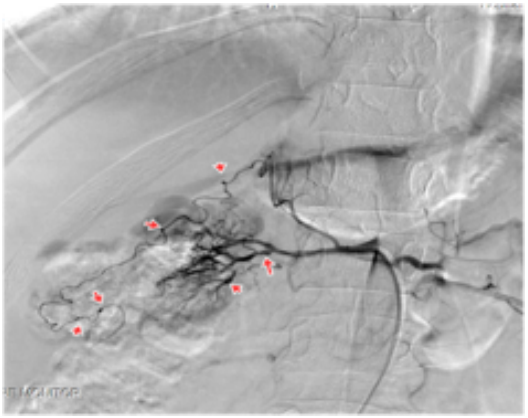

figure 5 : selective angiography showing aneurysmal dilatations of the right renal artery.

Thus, our patient had a perforated ulcer, vascular involvement and neurological involvement. This association was suggestive of systemic vasculitis. Erythrocyte sedimentation rate was of 110mm / hour. Anti-Neutrophilic Cytoplasmic Autoantibody (ANCA), serum cryoglobulins and anti-double-stranded DNA antibody (anti-dsDNA) were negative. Complement levels were within normal ranges. Finally, in view of the negativity of the ANCAs and the absence of glomerular or respiratory involvement, the diagnosis of PAN was made. The disease was not associated with hepatitis B. In addition to the treatment of hypertension which

was based on nicardipine, therapeutic management combined corticosteroids and cyclophophamide that was maintained for 18 months. Blood pressure was rapidly normalized. Two months later, the inflammatory syndrome regressed.

DISCUSSION :

PAN is a rare multisystemic vasculitis syndrome characterized by necrotizing arteritis of small and medium-sized arteries (1). It is rare in childhood. The pathophysiology is still unclear and it is likely that environmental triggers and host susceptibility play a key role. Otherwise, the intervention of infectious agents in the pathogenesis of PAN is supported by the frequent association of hepatitis B and other viruses such as HIV, hepatitis C, CMV and parvovirus B 19 with PAN (3). Because of its scarcity, the epidemiology of PAN remains indefinite. However, the onset of the disease is typically between the ages of 40 and 60 years (2). The age of revelation represents a peculiarity of the presented case even if some pediatric cases have been reported previously (4). In fact, of the primary vasculitis, Henoch Schönlein purpura and Kawasaki disease are the most common in youth and childhood PAN remains rare.

The clinical presentation is variable and broad. It is often misleading especially in early onset PAN. Constitutional symptoms, such as prolonged fever, and weight loss, are the most frequent but they were absent in the presented case. In addition to general symptoms, PAN may affect any organ system but the most commonly affected sites are the skin, joints, kidneys, gastrointestinal tract, and peripheral nerves (5). Cutaneous involvement includes usually purpura, livedo, subcutaneous nodules, and limb edema. It may be the only clinical manifestation in cutaneous forms without systemic involvement (6). The absence of cutaneous involvement in our patient made the diagnosis more difficult. Gastrointestinal involvement occurs in %14 to %65 of patients with PAN and is a major cause of morbidity and mortality (7 ,5). Abdominal pain is the most common symptom and small bowel and gallbladder are most commonly affected. Digestive involvement includes ulcerations, bleeding and transmural ischemia which may lead to the necrosis of the bowel wall with increased risk of perforation (7). In the presented case, the digestive involvement which revealed the disease was a duodenal ulcer without other locations.

The most common neurological manifestation of PAN is mononeuritis multiplex caused by vasculitis of the vasa nervosum (8). The involvement of the central nervous system is rare. Our patient combined peripheral neuropathy and central involvement. The PRES syndrome was likely secondary to nephrogenic hypertension. Although the mechanism of PRES syndrome is not clearly understood, hypertension associated with failed autoregulation and hyperperfusion are considered possible mechanisms (9).

Because of the lack of specific signs or serologic tests, the diagnosis of PAN is rather difficult. However, angiography is very helpful for diagnosing vasculitis, but the angiographic findings are not specific for PAN (11 ,10). In the presented case, the diagnosis was made on the basis of the association of digestive, neurologic and renal involvement and the presence of renal artery aneurysms suggesting a vasculitis. On the other hand, the absence of alveolar hemorrhage or glomerular involvement, the negativity of ANCA and serum cryoglobulins allowed the exclusion of differential diagnosis such as microscopic polyangiitis, mixed cryoglobulinemic vasculitis and ANCA associated vasculitis (12). The management of PAN is based on the Five-Factor Score (13). This score is useful to predict prognosis as well as guide treatment strategies. For patients with FFS of 1 or greater, immunosuppressive agents are associated with prednisone (14 ,13). Our patient combined digestive and neurological involvement. He had a score of 2 and thus had a poor prognosis in theory. However, he had good outcome. The peculiarities of our case were multiple. Firstly, the early onset of the disease represents a diagnostic difficulty view the rarity of this entity in childhood. Secondly, the disease was revealed by an initially isolated digestive tract involvement. Isolated duodenal localization during digestive tract involvement of PAN is rare. This condition mimed ulcerative disease although peptic ulcer is unusual at this age. Moreover, our patient combined central nervous involvement and peripheral neuropathy. The PRES syndrome reported in this case is also rare.

CONCLUSION :

Polyarteritis nodosa can present with serious symptoms such as profuse gastrointestinal bleeding.

Gastric and duodenal localizations of the gastrointestinal involvement of PAN are rare.

However, a high index of suspicion is necessary when dealing with seemingly obscure digestive bleeding. In these cases, precocious investigations including at least the erythrocyte sedimentation rate should be performed. In fact, early diagnosis is imperative to avoid the occurrence of other manifestations and to improve the prognosis.

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Contributors : all authors provided care for the patient, Sonia Mazigh and Samir Boukthir supervised the work.

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