Unusual Complications after a Severe Self-Limited Hemolytic Anemia — A Case Report

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Abstract

Auto-immune haemolytic anemias (AIHA) are a group of heterogeneous diseases of acquired immunologic destruction of red blood cells (RBC). Cold AIHA is usually associated with underlying disorders such as infections, autoimmune diseases and neoplastic growth. Infection associated cold AIHA may be severe, but is usually a self-limited disease without chronic complications of hemolysis such as gallstones or nephrolithiasis. We present a case of AIHA with auto-limited production of cold agglutinins associated with an acute infection and several uncommon lithiasic complications that occurred months after resolution of the hemolytic event.

ABBREVIATIONS

AIHA: Auto-Immune Hemolytic Anemia; RBC: Red Blood Cell; LDH: Lactate Dehydrogenase; CT: Computed Tomographic; ED: Emergency Department

INTRODUCTION

Hemolytic anemias are characterized by premature destruction of red blood cells (RBCs) and are usually classified in intrinsic (if the mechanism is intrinsic to the RBC such as hereditary membrane and enzymatic defects or hemoglobinopathies) or extrinsic/secondary (such as microangiopathic hemolytic anemias, autoimmune hemolytic anemias or infections of RBCs). The first tend to have a more indolent nature, with chronic hemolysis and acute hemolytic crises, while the latter are usually more acute and self-resolving [1].

In autoimmune hemolytic anemias (AIHAs) the autoantibodies target red blood cell (RBC) surface antigens, resulting in their destruction [2,3]. Depending on the optimum RBC-autoantibody reaction temperature, the AIHAs are divided into warm (if the optimum temperature is 37°C), cold (if it is less than 37°C) or mixed subtypes, and are further classified into primary or secondary in nature. [2] Most cold AIHAs are secondary and are associated with other disorders including autoimmune diseases, malignancies and infections [3-6].

CASE PRESENTATION

A 37 year old man, immune competent, was admitted because of a severe auto-immune hemolytic anemia, due to cold agglutinins, that appeared three weeks after a self-limited episode of abdominal pain and aqueous diarrhea without blood or mucus, which the patient interpreted as viral gastroenteritis and resolved without medical assistance. He maintained severe hemolysis without response to heating or methylprednisolone pulses. He was administered immunoglobulins and had 8 sessions of plasmapheresis with subsequent positive and sustained response with progressive reduction of markers of hemolysis. All study of neoplasms, including lympho proliferative diseases, and immune diseases was negative. A computed tomographic (CT) scan of the abdomen was performed that revealed the presence of splenomegaly (spleen with 16.5cm), without other alterations.

He had chronic gastritis, for which he was medicated with esomeprazole. He had no other known diseases and took no other medications. He had no history of allergies. He was married, worked as a driver in his brother’s wood company. He drank alcohol in moderation, did not smoke or use illicit drugs, and was physically active. There was no familiar history of significant diseases or health problems.

He was discharged after 14 days of hospitalization without immunosuppressive therapy. Five weeks after discharge, he returned to the emergency department (ED) with severe pain in the epigastrum. On physical examination, he had tenderness in the right upper abdominal quadrant, without other alterations. Complete blood count was normal and other laboratory values including hepatic transaminases were normal and there was no evidence of hemolysis. He was diagnosed with lithiasic

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- Gallstones
- Nephrolithiasis
- Spontaneous rupture of the urinary collecting system
cholecystitis, with abdominal echography and CT showing a non-distended gallbladder with normal wall dimensions but slight edema, with biliary sludge that was not present five weeks earlier and there was also evidence of microlithiasis on the left renal pelvis de novo (Figure 1). The patient received a 7 day antibiotic course of piperacillin/tazobactam and was discharged 7 days after admission without symptoms.

About 36h after discharge, he returned to the ED with acute severe left lumbar pain, constant, associated with hematuria and dysuria. He had no history of trauma. An abdominal CT showed a rupture of the left renal pelvis with leakage of contrast product to the peritoneum, without evidence of renal lithiasis (including the microlithiasis on the left renal pelvis) or obstruction (Figure 1). Retrograde ureteral catheterization was performed with placement of a double J stent. He evolved favorably, with symptomatic and imagiological resolution. He remained without evidence of hemolysis. Other entities such as vasculitis (such as polyarteritis nodosa) were excluded, as he had a normal thorax and abdominal angiography without any evidence of alterations including aneurysms. The patient was discharged at day 13 after admission, oriented to the urology clinic to follow up.

So far, ten months after initial admission he evolved favorably without further intercurrences.

**DISCUSSION**

The infection associated cold AIHA may require aggressive treatment in patients critically ill or with severe hemolysis, including corticosteroid therapy, plasmapheresis and erythrocyte transfusions, has occurred with our patient[3,4,5,7]. It is usually a self-limited event with good prognosis after infection resolution [3,6].

The destruction of RBCs leads to serum hyperbilirubinemia, and prolonged hemolysis may lead to the precipitation of bilirrubin salts (mostly calcium bilirubinate) and formation of pigmented stones within the gallbladder or biliary tree that, like any other gallstone, may lead to acute or chronic cholecystitis, biliary obstruction, or any other biliary tract consequence of calculous disease[8]. This is a well-established complication of chronic acquired or hereditary hemolytic anemia and the British Society of Haematology recommends prophylactic cholecystectomy[9]. We could not find in the literature any description of such occurrence after an acute, self-limited event. CT scan on the first admission excludes its existence previously to hemolysis.

Rupture of the urinary collecting system associated with peripelvic extravasation of the urine is an unusual condition and commonly associated with, although rarer causes include neoplasms, trauma, and iatrogenic procedure can occur [10].

The existence of microlithiasis in the affected side and the absence of trauma make obstructive calculus the most plausible cause of rupture in our patient. Non identification of calculus in the time of the rupture might be because of its elimination (since the beginning of the complaint until the diagnosis 12 hours passed). Conditions associated with high cell turn-over, such as hemolytic anemias, polycythemia vera and sickle cell disease, appear to be associated with higher incidence of uric acid nephrolithiasis probably related to the higher blood levels of uric acid [11]. However, this kind of calculus are also usually associated with chronic diseases and hemolysis [12,13].

Due to the severity of these complications, especially of spontaneous rupture of renal pelvis, the clinics must be aware of this possibility even in cases of self-limited hemolytic events.
REFERENCES