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An unusual case of autoimmune hemolytic anemia-thinking outside the box

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Case Report: An Unusual Case of Autoimmune Hemolytic Anemia- Thinking Outside the Box

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Introduction

Warm autoimmune hemolytic anemia (WAIHA) can be a life threatening entity if complicated by hypersplenism and reticulocytopenia. Overall, this is an infrequent complication. Here we present a case of WAIHA showing a drastic response to partial splenic artery embolization (PSAE).

Hospital Course

A 27 year old male with no significant past medical history presented to an outside hospital for chief complaints of nausea/vomiting, epigastric pain, and acute jaundice. Initial laboratory workup revealed a hemoglobin of 5.5 g/dl, normal WBC and platelet count, elevated direct bilirubin (10 mg/dl), elevated lactate dehydrogenase (969 IU/L), low haptoglobin (<8 mg/dl), the peripheral smear revealed rare polychromasia and spherocytes, a positive direct antiglobulin test (DAT: BS +, IgG +, C3d -) with presence of warm autoantibodies. All infectious disease testing was negative. Anti nuclear antibodies were not identified. The initial CT abdomen showed a normal sized spleen. The patient was diagnosed with idiopathic warm autoimmune hemolytic anemia and was treated with pulsed steroids, one dose of Rituximab, and transfused with a total of 13 units packed red blood cells. The patient did not respond to medical treatment, and after one week, the patient was transferred to our institution for escalation of care.

At our institution, Transfusion Medicine was consulted and advised only transfusion of phenotypically matched pack red blood cells for significantly symptomatic anemia or hemodynamic instability, as the patient continued a downtrend in the hemoglobin despite continued transfusion support. Decadron was added to his therapy, as well as IVIG that needed to be discontinued due to infusion related shortness of breath and chest pain. Another dose of Rituxan along with vincristine and cyclophosphamide were also given. Despite these efforts, the patient's hemoglobin continued to drop despite an additional 9 RBC units transfused at our institution, reaching a low of 2.1 g/dl. In addition, the patient developed left lower quadrant pain accompanied by bloody bowel movements. A repeat CT scan of the abdomen was performed, which revealed an enlarged spleen with a span of 16.5 cm in its maximum dimension. As the patient was not a candidate for splenectomy, partial splenic artery embolization (PSAE) was performed.

The lower to mid portions of the spleen were embolized individually with 500-700 micrometer Embosphere microspheres followed with a gelfoam/ contrast slurry mixed with gentamicin. Thirty percent residual viable spleen was seen on the final digital subtraction angiography (DSA). Six hours post procedure, a dramatic increase in hemoglobin was observed without any additional transfusion support. The patient suffered severe left upper quadrant abdominal pain that was controlled medically. After close observation, the patient was discharged home 2 days later. At home the patient was doing well symptomatically with the exception of persistent left upper quadrant pain. He had no clinical or laboratory signs of recurrent hemolysis. Hemoglobin was 12.1 g/dl. 3 months after embolization, patient underwent laparoscopic splenectomy due to persistent left upper quadrant pain. During this hospitalization a type and screen was performed in preparation for surgery. No warm autoantibodies were detected; however, additional new alloantibodies were identified.

Tables

Laboratory Markers for Hemolysis

Time frame	HGB (g/dL)	PLT (K/uL)	Retic (%/A)	HTG mg/dl	LDH IU/L	TB mg/dl	Lactate mmol/L
At admission	4.5 to 2.1	132	0.7(9.3)	< 30	880	11.8	NP
Pre embolization	2.1 to 4.5*	45	NP	< 30	ND	4.7	3.9 to 2.6*
Post embolization	5.7 to 6.7	88	1.9 (36.6)	< 30	1461	3.7	1.6
At discharge	6.6	345	2.2 (42.1)	214.7	489	1	ND
4 months post discharge	13.6	588	NP	NP	NP	0.2	0.9

* RBC transfusions received; HGB (Hemoglobin), Retic (%/Absolute Count), LDH (Lactate Dehydrogenase), HTG (Haptoglobin), TB (Total Bilirubin); NP (not performed)

Immunohematology Testing

Time frame	DAT (BS/IgG/C3d)	Eluate	Ab Screen	Ab Identification
At admission	2/2/0	negative	positive	WAA, allo anti-E
4 months post discharge	0/0/0	Not indicated	Positive	allo anti-E, anti-c, anti-K

Images

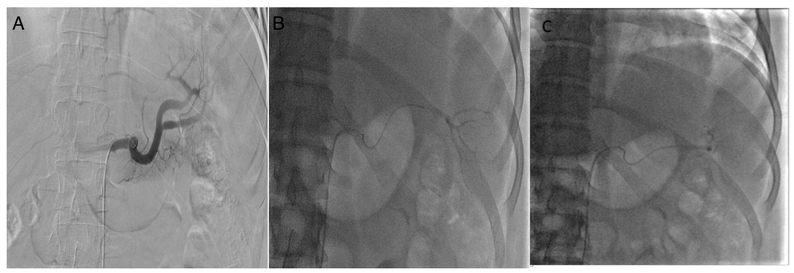

Figure 1: A) Digital subtraction angiography (DSA) at the proximal splenic artery B) DSA showing the lower splenic pole branches being selected C) DSA after embolization showing lack of opacification of the lower pole branches.

Discussion

Hypersplenism and/or reticulocytopenia are infrequent complications of a warm autoimmune hemolytic anemia, constituting a life threatening complication that may not respond to standard therapy and may not qualify for surgical intervention[1]. The above patient did not improve with any medical therapies, continued with a decreasing trend in hemoglobin that was accompanied by increasing lactate levels with no elevated troponin or signs of cardiac compromise. PSAE achieved a dramatic response that was maintained through follow up. Different embolic agents can be used including gelatin sponge pledgets, coils, plugs and microspheres. Techniques between different centers varies greatly but the Spigos technique has been shown to reduce complications[2].

Potential complications that can result from the procedure include pleural effusion, pancreatitis, paralytic ileus, or post-embolization syndrome[2]. Post-embolization syndrome can consist of leukocytosis, fever, and continued abdominal pain[3]. Due to recurrent pain from the embolization, our patient eventually received a splenectomy once he was stable.

In summary, we show that partial splenic artery embolization can be a safe and effective therapy for managing medication refractory warm autoimmune hemolytic anemia in patients who are at high risk for surgery.

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