Complicated Henoch-Schönlein Purpura And Antiphospholipid Syndrome: A Rare Association

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Citation

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Abstract

Henoch- Schönlein purpura (HSP) is the most common vasculitis in pediatric patients, with a small vessel vasculitis characterized by palpable purpura, arthritis, renal and gastrointestinal involvement.
We present the case of a child, showing signs of Henoch– Schönlein purpura, who developed an acute scrotum and some days later an acute abdomen.
He presented a prolonged activated partial thromboplastin time (aPTT) and Antiphospholipid antibody tests were positive for lupus anticoagulant antibodies.
This suggests that an antiphospholipid syndrome should be considered in cases of Henoch– Schönlein purpura and antiphospholipid antibodies should be measured to determine whether prophylactic antithrombotic treatment is needed to prevent thrombotic manifestations.

INTRODUCTION

Henoch- Schönlein purpura (HSP) is the most common vasculitis that occurs in pediatric patients, with a multi-organ involvement ([1]). Typically skin (with a characteristic rash present in all patients) and joints of arms and legs, gastrointestinal tract, kidneys are affected ([2];[3]). Some males also present external genitalia involvement ([4]) and/or acute scrotum, with skin petechiae of buttock, legs and infrequently of the upper torso and extremities. Most patients have a good prognostic outcome, with resolution without sequelae. However sometimes severe complications occur, as nephritis or renal failure, intestinal occlusion, lung hemorrhage, CNS compromission with headache, stroke or palsy, death.

The antiphospholipid syndrome is the most common acquired thrombophilia of autoimmune etiology and may occur as a primary clinical entity or in association with an underlying systemic disease as Systemic Erythematosus Lupus. It is characterized by arterial and venous thrombosis with various clinical manifestations in the presence of antiphospholipid antibodies ([5]). We describe a four-years old boy with a rare association of HSP with antiphospholipid syndrome.

CASE REPORT

In our division a four years-old boy was admitted for the occurrence, four days before the admission, of arthralgia of the right leg, treated at home with Ibuprofen with symptomatic improvement. The day before the admission he presented purpura of the skin of the right ankle, of the bottom and of the knees. At the admission to our clinic the boy was sufferer, with skin palpable purpura of the legs, generalized abdominal pain and acute scrotal pain with swelling and scrotal skin petechiae.

Laboratory examinations showed: total leukocyte count increased (11.620/µl); platelet count, hemoglobin, transaminases, total proteins and albumin, RCP, ESR, C3, C4, IgA, IgM, IgG, fibrinogen, aPTT, PT: in the normal range; microhematuria. The scrotal sonographic imaging evidenced increased volume of both epididymis with reactive hydrocele and scrotal wall thickening. He also made an abdominal ultrasonography that showed diffuse intestinal meteorism.

The surgeon supported the diagnosis of HSP related epididymitis. He started therapy with methylprednisolone (2 mg/kg/die). The patient promptly improved with the resolution of his acute scrotum and the decrease of articular swelling and of purpura.

Two days after the recovery he was admitted in a surgery unit, showing an acute and generalized abdominal pain with new skin rash. A direct radiography showed diffuse bowel

distension, without signs of intestinal occlusion or perforation, as confirmed by ultrasound.

He started treatment with methylprednisone (2 mg/kg//die) and ceftriaxone (100 mg/kg/die) with a rapid resolution of his abdominal involvement. Hence he was transferred back to our unit.

In retested detections during the follow up (see table 1) a coagulation evaluation revealed a prolongation of activated partial thromboplastin time (aPTT: 106 sec; cut off 20-40 sec). Hence coagulation factors were studied: the patient had reduced levels of factor VIII and of its inhibitor; we also studied autoimmunity to exclude SLE. He showed normal values of ANA, ENA, anti-dsDNA, ANCA, anti beta 2 glycoprotein I antibodies with the positivity of Lupus Anticoagulant.

He continued the treatment with scalar doses of corticosteroids and at the present time he is asymptomatic and followed in our unit every month to monitor clinical outcome, autoimmunity pattern and coagulation tests.

After two months he presented anticardiolipin antibodies IgM positive (31.9 MPL/ml; normal range: 0.0-12.0), with anticardiolipin antibodies IgG negative (11.7 GPL/ml; normal range: 0.0-12.0).

Figure 1

Table 1: aPTT levels detected in the patient

	02/11/07	10/11/07	10/11/07	12/11/07	16/11/07	19/11/07	24/11/07	19/12/07
aPTT (sec)	28,8	106	94	115	103	89,4	75,1	54,9

DISCUSSION

HPS is an autoimmune acute leukocytoclastic vasculitis more frequent in children between 4 and 11 years of age and has an incidence of 14 in 100.000 population.

It is characterized by deposition of immune complexes as a response to infections, drugs, etcetera.

HSP is typically characterized by palpable non-thrombocytopenic purpura on initial clinical presentation. Articular, renal and gastrointestinal involvement could be associated.

The common GI features are abdominal pain (58%), massive colorectal bleeding or occult blood loss (20%) ([6]). Major complications of abdominal involvement develop in 4.6% (range 1.3-13.6%), of which intestinal perforation, intussusception, and infarction constitute the major surgical complications in HSP. Ultrasonography complements clinical assessment, clarifies the nature of the gastrointestinal involvement and reduces the likelihood of unnecessary laparotomy ([7]).

Early corticosteroid treatment for intestinal complications is recommended.

Epididymitis is an inflammatory process generally affecting males from 9 to 14 years of age, however it can be seen in younger males with Kawasaki disease and Henoch-Schönlein purpura ([8]), as in our patient.

Complicated SHP is a clinical condition that need accurate follow up and a complete diagnostic pattern to exclude associated diseases as systemic lupus erythematosus or other rheumatic processes as antiphospholipid syndrome. Serum levels of IgA anticardiolipin antibodies (aCL) are elevated in the initial active stage of adult HSP, suggesting that the IgA aCL may play some role in the onset of adult HSP ([9]).

An association between antiphospholipid syndrome (APS) and HSP is reported, especially in complicated cases ([5];[10]). The APS is the most common acquired state of hypercoagulation both in adults and in children. The autoimmunity is supposed to be triggered by environmental factors (infections, drugs, traumatic injuries) in genetically predisposed patients. Two forms of APS are described: the primary form and the secondary one, associated with other diseases, first of all Systemic Lupus Erythematosus (SLE) ([5]). In children APS commonly occurs as arterial or venous thrombosis and rarely in association with neurological or hematological signs and symptoms, otherwise frequently described in adults, and with thrombocytopenia associated with serologic markers (anticardiolipin antibodies, anti beta 2 glycoprotein I antibodies and positive Lupus Anticoagulant test).

In APS skin involvement is often the first sign (livedo reticularis, ulcerations, digital gangrene, subungueal splinter hemorrhages, superficial venous thrombosis, thrombocytopenic purpura, pseudovasculitic manifestations, extensive cutaneous necrosis, and primary anetoderma).

The progressive positivization of autoimmunity of this patient seems to be triggered by HSP or to be linked to the etiology of the purpura. Furthermore our child developed a complicated form of HSP, with abdominal symptoms mimicking an ischemic intestinal pattern.

Our case suggests that an APS should be considered in cases

of symptoms evocative of arterial or venous thrombosis, or in cases of abnormalities of coagulation state in patients affected by HSP.

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