

# Henoch–Schönlein purpura (HSP) and high-dose immunoglobulin treatment in patient with familial prostatic adenocarcinoma

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A 52-y old man was admitted to our department because of abdominal pains and diarrhea with fresh blood, with concomitant purpura over the whole body and edema of the both tarsal joints. The medical history of the patient included skin changes of similar character identified once 10–12 y before. The family history revealed prostate cancer (brother and father) and pulmonary carcinoma (mother and mother's sister). An administration of immunoglobulins in the course of HSP is a non-standard clinical procedure and in case of our patient—clinically effective. In the literature, we have found only few articles about intravenous immunoglobulin treatment for acute, adult-onset HSP and only one article about GI bleeding from colonic ecchymoses in HSP. In these cases HSP wasn't associated with prostate cancer. In the first article, authors had seen dramatic responses to intravenous immunoglobulin, like in the case presented by us. IV-IG acts as an immunomodulator by suppression of antibody production, Fc-receptor blockade and anti-idiotypic reaction. In our case, the last two mechanisms could be perceived as favorable effects of IV-IG.

A 52-y old man was admitted to our Department because of abdominal pains and diarrhea with fresh blood, with concomitant purpura over the whole body and edema of the both tarsal joints. The medical history of the patient included skin changes of similar character identified once 10–12 y before. The family history revealed prostate cancer (brother and father) and pulmonary carcinoma (mother and mother's sister).

On admission, laboratory assays provided normal coagulation parameters—APTT, INR, elevated fibrinogen and D-dimers, decreased antithrombin III levels, increased glucose and HBA1C (9,2) levels and elevated PSA (>150).

Preliminary diagnosis: Bleeding from the lower gastro-intestinal tract; suspected malignancy of the large bowel and of the prostate gland, Henoch–Schönlein purpura (HSP) and first diagnosed type 2 diabetes mellitus.

Insulin and steroid administration completed the mainline treatment.

In the course of hospital observation, purpura progressed to develop hemorrhagic bullae, including distal parts of the limbs, and periodical symptoms of acute abdomen occurred. Laboratory tests—including blood count—revealed: CRP up to 68.63, slight anemia (Hb 10–13 g/L, thrombocythaemia to  $565 \times 10^3/\mu\text{L}$ , periodical leucocytosis of  $10\text{--}20 \times 10^3/\mu\text{L}$ , negative pANCA and pANA values, IgA and IgE within standard values, decreased IgG and IgM levels; in proteinogram: decreased immunoglobulin levels, normal C3 value and increased C4 level, normal hepatic

parameters, negative blood and urine cultures, while proteinuria and erythrocyturia occurred in the course of hospitalization.

Colonoscopy of the terminal section of the small intestine revealed ulceration with active bleeding and sigmoid diverticulosis. Histopathological results included “Enteritis chronica activa cum ulceratione, tela neoplasmatica absenta” (Chronic enteritis with ulcerations, no presence of neoplastic tissues). CT imaging of the abdominal cavity demonstrated a visible, continually thickened, oedematous wall of the ileum with an excessively enhanced contrast of the mucous membrane—suggestive of inflammatory changes, and oedematous changes in the mesoileum. No tumor was identified. A follow-up CT, performed after two weeks, presented a considerably lower range of oedematous changes within the intestines. Currently, there is a segmental thickening with wall edema in the descending part of the duodenum, in a short section of one of the jejunal loops, as well the cecum and the ascending colon, while the oedematous walls of the ileum, visible in previous imaging, now is only slightly thickened and on a short section with improved mesentery image.

An additional, precise, thin-layer thoracic CT revealed no pathologies, except slight fluid volumes in both pleural cavities.

Insulin and steroid therapies were continued, furthermore a broad-spectrum antibiotic therapy was added after the first CT imaging. Facing a weak response to the applied treatment with deterioration of the general condition of the patient (growing weakness, fever, progression of skin changes and joint pains

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plus maintained diarrhea with blood addition) and low immunoglobulin levels in blood, a decision was made to administer intravenously (i.v.) immunoglobulins (IV-IG)—on the 25th day of hospitalization, a single administration of 20 g of Octagam (human immunoglobulin), 4 times daily a 5-g ampoule of OCTOPHARMA LTD—followed by an evident improvement in the patient's well-being (within six days, abdominal problems and diarrhea regressed with almost total regression of skin changes, CRP dropped down to 3.6; thrombocythaemia was up to  $538 \times 10^3/uL$ , and leukocytosis were maintained; the other, above-mentioned parameters, were not controlled during hospitalization before immunoglobulin administration).

Biopsy of the prostatic gland was performed with diagnosis of adenocarcinoma.

The hospitalized patient was consulted by hematologist, rheumatologist, dermatologist and surgeon.

The entire clinical presentation of the described patient suggests the paraneoplastic syndrome in the course of familial prostate carcinoma with positive family history of the condition as well with history of pulmonary neoplasms and type 2 diabetes mellitus—what, accordingly, predisposes to the described changes, typical for HSP. The observed intestinal changes should be treated as HSP symptoms. An administration of immunoglobulins in the course of HSP is a non-standard clinical procedure and in case of our patient—clinically effective.

Diagnosis: Prostate carcinoma–adenocarcinoma (familiar type). Leukoclastic vasculitis (Henoch–Schönlein purpura with normal IgA plasma level) in the course of primary condition. Intestinal and gastric mucositis, renal damage and arthritis in

the course of vascular changes. Insulin-controlled type 2 diabetes mellitus. Sigmoid diverticulosis. Gastresophageal reflux with oesophagitis. Hiatus hernia.

The patient was referred to the urological center to be qualified for further treatment.

Case study justification:

- family history of prostate cancer in a patient with family history of pulmonary neoplasms and type 2 diabetes mellitus;
- typical for HSP, vascular changes (leukoclastic vasculitis in the course of primary disease, supported by dermatologic counselling) with significant gastric tract involvement;
- a rapid turn after the i.v. immunoglobulin administration toward considerable improvement of the patient's general well-being, enabling further instituting of diagnostic and therapeutic protocols.

In the literature, we have found only few articles about intravenous immunoglobulin treatment for acute, adult-onset HSP<sup>1,2</sup> and only one paper about GI bleeding from colonic ecchymoses in HSP.<sup>3</sup> In these cases HSP wasn't associated with prostate cancer. In the first article authors had seen dramatic responses to intravenous immunoglobulin, like in the case presented by us. IV-IG acts as an immunomodulator by suppression of antibody production, Fc-receptor blockade and anti-idiotypic reaction.<sup>4</sup> In our case, the last two mechanisms could be perceived as favorable effects of IV-IG.

#### Disclosure of Potential Conflicts of Interest

No potential conflicts of interest were disclosed.

#### References

1. Raniele DP, Opsahl JA, Kjellstrand CM. Should intravenous immunoglobulin G be first-line treatment for acute thrombotic thrombocytopenic purpura? Case report and review of the literature. *Am J Kidney Dis* 1991; 18:264-8; PMID:1867184
2. Kusuda A, Migita K, Tsuboi M, Degawa M, Matsuoka N, Tominaga M, Kawakami A, Kawabe Y, Taguchi T, Eguchi K. Successful treatment of adult-onset Henoch-Schönlein purpura nephritis with high-dose immunoglobulins. *Intern Med* 1999; 38:376-9; PMID:10361914; <http://dx.doi.org/10.2169/internalmedicine.38.376>
3. Cappell MS, Onea M, Amin M. GI bleeding from colonic ecchymoses associated with cutaneous ecchymoses over the lower extremities from leukocytoclastic vasculitis mediated by IgA deposition in Henoch-Schönlein purpura. *Dig Dis Sci* 2011; 56:1919-20; <http://dx.doi.org/10.1007/s10620-010-1502-0>; PMID:21136164
4. Dwyer JM. Manipulating the immune system with immune globulin. *N Engl J Med* 1992; 326:107-16; PMID:1727218; <http://dx.doi.org/10.1056/NEJM199201093260206>