

Ulcerative colitis accompanied by aseptic abscesses syndrome and small-vessel vasculitis

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Abstract

Introduction. Cutaneous small-vessel vasculitis and aseptic abscesses syndrome are among the rarest extraintestinal manifestations of bowel inflammatory diseases.

Materials and methods. The study describes the case of a 19-year-old female admitted to the Department of Dermatology due to cutaneous small-vessel vasculitis. In order to reach the final diagnosis, a wide spectrum of laboratory and diagnostic tests were performed. Three subsequent abdominal CT scans confirmed the presence of hypodense foci, initially in the spleen, and subsequently in the liver. The aseptic abscesses syndrome was diagnosed based on the radiological picture and the exclusion of infectious causes of the above-described foci.

Objective. Presentation of a case of concomitance of cutaneous small vessel vasculitis and septic abscess, accompanied by inflammatory bowel disease.

Results. During the hospitalisation, the patient developed symptoms of paralytic ileus. The results of colonoscopy and biopsy of intestinal mucous membrane were typical of ulcerative colitis.

Conclusions. To the best of the authors' knowledge, this is the first presentation of ulcerative colitis accompanied by aseptic abscesses syndrome concomitant with cutaneous small vessel vasculitis.

Key words

leukocytoclastic vasculitis, aseptic abscesses, inflammatory bowel disease

INTRODUCTION

Vasculitis forms a heterogeneous group of diseases characterised by primary involvement of vascular structures by an inflammatory process causing damage to vascular walls, and in the case of disrupted blood supply to body organs – to their dysfunction [1, 2, 3, 4].

Leukocytoclastic or hyperergic vasculitis (LcV) is the most common form of cutaneous small-vessel vasculitis [5, 6, 7]. In 45–55% of cases the cause of the vasculitis remains unknown, in 15–20% of patients it is preceded by an (bacterial, fungal or viral) infection, in 15–20% connective tissue disorders or inflammatory bowel diseases (IBD) are the causative factor, 10–15% is triggered by drugs, while neoplasms are responsible for approximately 5% of cases [8, 9, 10].

The aseptic abscesses syndrome (AA) belongs to a group of autoinflammatory diseases. It is characterised by the presence of deep abscesses, which are the consequence of accumulation of neutrophils, detected on radiological tests, negative results of blood and abscess aspirate cultures, and no response to antimicrobial agents and rapid post-glucocorticosteroid (GCS) improvement [11, 12, 13, 14].

In the majority of described cases, aseptic abscesses occur in the spleen (70%), lymph nodes of the abdominal cavity (53%), liver (40%), lungs (17%), pancreas (10%), brain (10%),

and sometimes also in the soft tissues. A relationship between the AA syndrome and the inflammatory bowel diseases, and neutrophilic dermatoses has been described [11, 12].

OBJECTIVE

The aim of this study is to present the case of concomitance of cutaneous small-vessel vasculitis and aseptic abscesses syndrome, accompanied by inflammatory bowel disease.

CASE STUDY

A 19-year-old female patient was admitted to the Department of Dermatology because of pleomorphic skin eruption consisting of indurative erythema, blisters, tumours and ulcerating tumours located on the lower legs, accompanied by joint contractures and oedema (Fig.1a-d).

Since 2008, the patient has been hospitalised several times, diagnosed with pseudomembranous enterocolitis (2008), erythema nodosum (2009), and Weber-Christian disease (2010). During each hospitalization, laboratory tests showed: leucocytosis – 15.8 K/μl, anaemia (Hgb) – 9.3 g/dl, RBC – 3.86 M/μl, thrombocythaemia (PLT) – 678 K/μl, neutrophilia (Neu) – 8.06 K/μl, (Neu) – 76.8%, increased CRP – 81.8 mg/l, and ESR – 58/h.

A histopathological examination of the lesional skin biopsy revealed a picture indicative of leukocytoclastic vasculitis, which was further confirmed by direct immunofluorescence

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Figure 1.

of the collected sample showing IgM immunoglobulin and C3 deposits in the vessel walls.

The patient underwent a number of diagnostic tests which excluded connective tissue diseases, infections and neoplasms. No infectious agents were identified either by mycological and bacterial cultures, nor by the Grocott and Ziehl-Neelsen staining of the biopsied lesions. Additionally, no infections were identified either in pantomographic rgt and throat cultures. A series of blood culture tests showed no bacterial growth; serological test results did not confirm an infection caused by *Borrelia burgdorferi*, *Chlamydia trachomatis*, *Treponema pallidum* or *Mycoplasma*.

To establish a differential diagnosis of bowel inflammatory lesions, tests for antibodies to *Yersinia enterocolica*, *Yersinia pseudotuberculosis* and Quantiferon tests were carried out. No parasites or *Clostridium difficile* toxins A and B were identified in a stool sample. Viral test results (antibodies to HIV, EBV, HCV, RNA-HCV, anti-HBs) were negative.

In order to exclude systemic connective tissue diseases and systemic vasculitis, the levels of anti-nuclear antibodies (ANA, ds-DNA, ASMA, LKM, pANCA, cANCA), anti-cardiolipin antibodies, cryoglobulins, antibodies to β 2-glycoprotein and C3, C4 complement components were determined. All the test results were within normal reference limits. The levels of Ca125, AFP, CEA tumour markers were also within normal limits.

In order to differentiate focal lesions in the liver, a blood serum test for tapeworm (Western-Blot) (*E. granulosus*, *E. multilocularis*), aspergillosis and systemic candidiasis (ELISA) were carried out with negative results.

Doppler ultrasound of lower-limb vessels showed no vascular pathologies or thrombosis.

Three months prior to the current hospitalization, the patient underwent an abdominal CT scan which showed the presence of hypodense lesions with a diameter of 15 mm, localised in the spleen (3 lesions) and the liver (2 lesions in segments 6 and 8) (Fig. 2a) which suggested metastatic lesions.

However, during the current period of hospitalisation, 2 abdominal CT scans were carried out with negative results for focal lesions in the spleen (Fig. 2b).

A hypodense focus in segment 5 of the liver was identified on the first CT scan (Fig. 2c). The next scan performed 7 days later, showed the same lesion, and additionally a lesion in segment 7 (Fig. 2d).

In the meantime, the patient experienced a sudden increase in body temperature – up to 39°C, symptoms of abdominal tympany, and bright red blood in per rectum examination. An abdominal X-ray was therefore carried out which showed symptoms of paralytic ileus (*megacolon toxicum*) – widening of and high fluid levels in the bowels. Abdominal ultrasound results showed fluid in the Morison's pouch. After the symptoms of paralytic ileus subsided, the patient underwent a colonoscopy. The colonoscopy results showed multiple ulcerations in the large intestine, with easy bleeding, covered with fibrin and mucosal oedema in the intestine. The lesions observed in the colonoscopy were classified in accordance with the Mayo Endoscopic Scoring of Ulcerative Colitis scale as Stage 3 lesions. The clinical presentation, endoscopic and histopathological test results

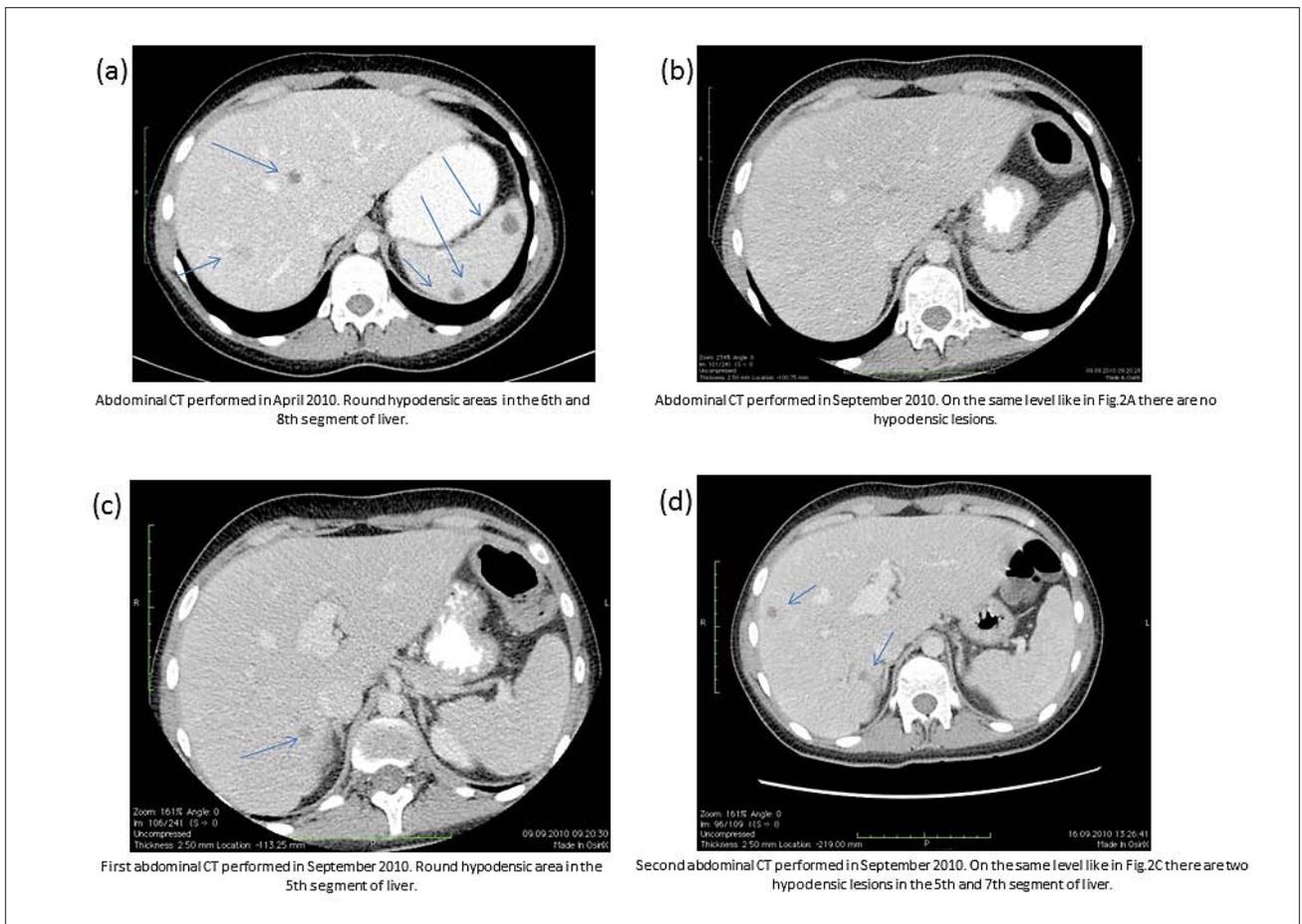


Figure 2.

were indicative of ulcerative colitis. The treatment included GCS, azathioprine and mesalazine, which resulted not only in resolution of intestinal symptoms, but also in near complete clearing of skin lesions.

DISCUSSION

To the best of the authors' knowledge, this is the first presentation of ulcerative colitis accompanied by aseptic abscesses syndrome, concomitant with cutaneous small-vessel vasculitis.

Chronicity and recurrence of skin eruptions in association with persistent anaemia, thrombocytopenia, elevated levels of inflammatory response markers suggested secondary nature of cutaneous lesions.

In the presented case, the diagnosis of cutaneous small-vessel vasculitis was confirmed by the results of histopathology and direct immunofluorescence (DIF) examinations [15, 16, 17, 18].

The diagnosis of ulcerative colitis was established based on the histopathological picture of the intestinal biopsy in connection with the typical course of the disease complicated by paralytic ileus.

The aseptic abscesses syndrome was diagnosed based on the overall clinical presentation, abdominal CT scans which showed significant dynamics of lesions according to imaging test results, concomitance of ulcerative colitis, laboratory test results – exclusion of infectious background (blood culture

test, serological tests for *Yersinia enterocolica*, Quantiferon), leucocytosis and elevated ESR.

The aseptic abscesses syndrome as described by Andre et al. [11, 12] is a group of clinical and pathological symptoms which include: young age, fever, abdominal pain, leucocytosis, elevated ESR. The imaging test results reveal hypodense, abscesses-like usually of multi-focal distribution, localised mostly in the spleen, liver, abdominal lymph nodes and sometimes outside the abdominal cavity, e.g. in the lungs, pancreas and brain [11, 12].

In 7% of cases lesions can be localised more superficially in the form of skin abscesses, sometimes with the involvement of superficial lymph nodes, muscles, pharynx or testicles. A proportion of affected patients will have also some articular complaints, such as arthritis and arthralgia. These symptoms do not respond to antimicrobial therapy, while regressing rapidly after GCS therapy or biologic agents [12, 19, 20].

The AA syndrome is accompanied by IBD in 66% of cases and by neutrophilic dermatoses in 20% of cases (e.g. Sweet's syndrome and pyoderma gangrenosum). There are also reports about concomitance of AA with rheumatoid arthritis or Weber-Christian disease [12, 18, 19, 20, 21]. The syndrome does not seem to be invariably accompanied by the conditions referred to above. Cases have been described in which the aseptic abscesses syndrome preceded or occurred subsequent to the above-listed disorders.

Andre et al. [12] suggested a number of diagnostic criteria for the aseptic abscesses syndrome (Tab. 1). The patient's

condition in the presented case complied with all these criteria.

The proper diagnosis of all the lesions was confirmed by the effective treatment of both bowel lesions and secondary skin lesions after administration of GCS and immunosuppressants.

CONCLUSIONS

Ulcerative colitis can be accompanied by aseptic abscesses syndrome and cutaneous small-vessel vasculitis; however, both the diagnosis and efficient treatment are difficult. It is therefore necessary to ensure the good collaboration of a number of specialists, as well as carrying out a wide spectrum of laboratory and imaging tests.

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