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Surgical Complications of Hemorrhagic Vasculitis in a Child

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Hemorrhagic vasculitis is a hematological disorder, which is often accompanied by abdominal pain syndrome and blood in stool, which is why it requires differential diagnosis with acute surgical pathology. The article presents clinical follow-up of a patient with hemorrhagic vasculitis complicated by intestinal intussusception and developed mesenteric thrombosis, which required emergency surgical intervention.

Keywords: hemorrhagic vasculitis, abdominal pain syndrome, intussusception, laparoscopy, mesenteric thrombosis.

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Introduction

Hemorrhagic vasculitis is one of a few hematologic diseases requiring surgical intervention in most cases [1-3]. It is necessary due to abdominal pain syndrome associated with punctate hemorrhagic rashes on the intestinal serosa and peritoneum [4]. The symptoms associated therewith require considering acute surgical disorders of abdominal organs for differential diagnosis. Moreover, due to the presence of stool blood in patients with hemorrhagic vasculitis, surgeons often have to consider intussusception for differential diagnosis as well [1, 3]. In most cases, although not always, diagnosis of intussusception in this category of patients is rather easy to rule out.

Below is a rare clinical case of follow-up and treatment of a child suffering from hemorrhagic vasculitis with developed intussusception and complicated by mesenteric thrombosis.

Clinical case

Child M., 13 years of age, was admitted to the Izmaylovo pediatric municipal clinical hospital (Moscow) with complaints of intense skin rash and pain in the left ankle joint when walking. Anamnesis shows that the child developed acute disease on June 06, 2012, when gradually progressing hemorrhagic rash appeared on lower limbs. The child was forwarded to the hematological center of the Izmaylovo pediatric municipal clinical hospital and later hospitalized after the diagnosis of hemorrhagic vasculitis had been established.

At admission: regularly colored skin, abundant maculopapular confluent hemorrhagic rash on lower limbs with exudation. Clear oropharynx. Left ankle joint – smoothed contours, movability intact. Vesicular pulmonary respiration, no rales. Clear and loud heart tones. Abdomen – not

swollen, soft, symmetrical on both sides, accessible to deep palpation in all segments, moderately tender around the epigastrium.

2 days after admission the child developed abdominal pain syndrome, which was terminated by means of infusion spasmolytic and anti-secretory therapy. Later, abdominal pain would recur at times.

The child underwent fibroesophagogastroduodenoscopy (June 13, 2012), which revealed multiple duodenal erosions. Intensity of the abdominal pain syndrome considerably decreased in the setting of antibacterial, anti-secretory, vasoprotective, anticoagulation and hormonal therapies of hemorrhagic vasculitis, as well as antiulcer therapy. Control fibroesophagogastroduodenoscopy (June 22, 2015) revealed positive changes (termination of erosive process in the duodenal bulb). However, a wave-like recurrence of skin manifestations in the form of punctate hemorrhagic rash continued to persist. The boy remained emotionally labile and had a rather low appetite, which is why the child refused sparing diet.

Laboratory tests. Complete blood count revealed leukocytosis up to 28 000 (normal range – $4-9 \times 10^9/l$), left (neutrophilic) shift to 85 (normal range – $18-77 \times 10^9/l$), ESR increase to 42 mm/hour (normal range – 2-20 mm/hour). Urine analysis (June 27, 2012) revealed microhematuria (8-10 per HPF; normal range – 0-3), proteinuria (1.0 g/l; normal range – 0-0.0002), persistence of recurrent skin hemorrhages, which is why the patient was prescribed glucocorticoid therapy.

Abdominal pain resumed on June 28, 2012; emesis with the eaten food appeared. The child visited a surgeon, who suspected intussusception. An upright radiographic examination of abdominal organs was performed; no free gas was found; however, multiple intestinal arches and liquid levels were observed in the upper segments. Loops of the small intestine were extended, lower segments of the abdominal cavity were obscured.

The child underwent pneumography; intussusceptum was detected and unfolded. The child received barium meal *per os*. 2 hours 40 minutes after barium intake the child underwent radiographic examination of abdominal organs to control intestinal passage. Contrast medium was observed in the stomach and in a small segment of the small intestine. Impossibility of ruling intestinal obstruction out, degradation of the child's condition and progression of the abdominal pain syndrome led to the decision to perform diagnostic laparoscopy.

Diagnostic laparoscopy. Laparoscopy revealed a large amount of opaque hemorrhagic exudate in the abdominal cavity and hyperemia of the parietal peritoneum. Intestinal loops were hyperextended and featured with fibrinous plaques. 2 perforations of the small intestine wherefrom intestinal contents entered the abdominal cavity were identified. The physicians in charge determined the need in lower midline laparotomy.

Revision at a distance of approximately 60 cm off the ileocecal junction demonstrated purple-bluish small intestine with necrotic patches and 3 perforations of 0.5-1.0 cm in diameter. Further revision revealed thrombosis of mesenteric vessels of the mesostenium. The altered loop of the small intestine underwent resection within the range of intact tissues. The total length of the removed part of the intestine was approximately 50 cm.

Double-barrel ileostomy was introduced by means of a laparotomy incision in the right mesogaster.

Biopsy of the small intestine: acute thrombosis, thromboarteritis of mesenteric vessels of the small intestine, acute hemorrhagic necrosis of a small intestine's segment, focal ulcerophlegmonous enteritis with perforation in the middle third of the small intestine's segment, fibrinopurulent mesenteriolysis, peritonitis.

The child was transferred from the operating room to the emergency department, where he started to receive a comprehensive glucose saline solution therapy and antibacterial therapy (III-generation cephalosporins and metronidazole, later – aminoglycosides, carbapenems), as well as replacement (albumin 10% [100 ml] and 20% [200 ml]), antifungal (fluconazole), anti-secretory (proton pump inhibitors), transfusion and enzyme (pancreatic enzymes) therapies. Coagulogram-controlled heparin therapy and antithrombin III injection were performed due to

manifested hypercoagulability events. Moreover, recurrent skin hemorrhagic syndrome and signs of nephritis (urinary syndrome) were observed in this period.

The boy's condition stabilized in the setting of the conducted treatment; the child was transferred to the department of surgery and, later, to the hematology unit to continue treating the background disease. The stoma functioned properly. Results of laboratory tests remained within the normal range, including coagulation parameters. The child remained at the hematology unit until October 08, 2012, when physicians succeeded in stabilizing the clinical pattern. On October 11, 2012, the patient's ileostomy was closed. Postoperative period was uneventful, skin hemorrhagic syndrome – none, urinalysis – no pathologies, coagulogram – within the normal range. The child was discharged in satisfactory condition to be followed up by a local hematologist, a nephrologist and a gastroenterologist.

Discussion

Hemorrhagic vasculitis was first described as anaphylactic purpura by German physician J.L. Schönlein, who published a treatise on it together with E.N. Henoch in 1874. In 1959, V.A. Nosova coined term “hemorrhagic vasculitis” in Russia [5]. Term “Henoch-Schönlein purpura” is still being used abroad.

Etiological factors of hemorrhagic vasculitis development may include various diseases, such as typhus, paratyphoid fever A and B, measles, yellow fever, although the most common trigger thereof is an upper respiratory tract disease [6]. Other trigger mechanisms of hemorrhagic vasculitis in children include drugs, especially antibiotics, food allergens, insect bites, rapid ambient temperature changes.

There are several classifications of hemorrhagic vasculitis.

Forms of the disease:

- cutaneous and cutaneo-articular: simple form, necrotic form and a form with cold urticaria and edemas;
- abdominal and cutaneo-abdominal;
- renal and cutaneo-renal (including a form with nephrotic syndrome);
- mixed [5].

In that case, the disease was of mixed nature. This conclusion is confirmed by the disease onset (the patient was admitted with complaints of intense skin rash [hemorrhagic, maculopapular]) and pain in the left ankle joint when walking. The child developed abdominal pain syndrome at the height of disease (2 days after hospitalization); it was terminated by means of infusion spasmolytic and other therapy. Later, abdominal pain would recur at times. Moreover, urinalysis demonstrated a range of changes (microhematuria and proteinuria).

One other classification of this disease is based on the rate of clinical development of hemorrhagic vasculitis.

There are the following types of the pathology course:

- fulminant;
- acute;
- subacute;
- prolonged;
- chronic [6].

In that case, the disease was of prolonged nature, as indicated by protracted resolution of clinical signs of hemorrhagic vasculitis: the first signs of the disease occurred in June 2012, while reconvalescence was achieved under rigorous follow-up and therapeutic monitoring only in November 2012.

There are 3 forms of hemorrhagic vasculitis in terms of the degree of activity:

- Degree of activity I – satisfactory condition of the patient, normal body temperature or low-grade fever, non-abundant skin rash, no other manifestations, high ESR (up to 20 mm/hour).
- Degree of activity II – moderately severe condition of the patient, body temperature over 38 °C (fever), manifested skin, intoxication (headache, asthenia, myalgiae) and articular syndromes, moderately manifested abdominal and urinary syndromes. Blood – high levels of leukocytes, neutrophils and eosinophils, high ESR (up to 20-40 mm/hour), low albumin level, traceable dysproteinemia.
- Degree of activity III is characterized by severe condition of the patient, evident symptoms of intoxication (high body temperature, headache, asthenia, myalgiae). Manifested skin, articular, abdominal (paroxysmal abdominal pain, vomiting blood) and renal syndromes. Moreover, central and peripheral nervous systems may be affected. Blood – significantly increased levels of leukocytes and neutrophils, high ESR (over 40 mm/hour); anemia and low platelet level may be observed.

Given this classification, we may conclude that the patient described in this article features pathology with degree of activity III.

According to the classification, clinical pattern varies depending on the form, course and activity of the process. Skin lesion is the most common symptom [3]. Characteristic hemorrhagic rash – the so called palpable purpura – is observed; its elements insignificantly rise above the skin surface; such a rise is not visible, yet it is easily identified on palpation. Separate elements often merge and may form continuous fields of significant area. Separate elements may (although rarely) necrotize. Rash may be petechial in the onset of the disease. In the onset of the disease the rash is always localized in distal segments of lower limbs. Later, it gradually spreads to hips and buttocks. Upper limbs, abdomen and back are only rarely affected [7]. After a few days purpura usually pales, becomes discolored to brown due to pigmentation and later gradually disappears. In the event of relapses, pigmentation sites may remain in place. Cicatrices are never observed (except for singular cases characterized by necrosis of elements and secondary infection overlay).

A characteristic feature of the articular syndrome is that it often develops together with skin syndrome; it is observed in 59-100% of cases [2, 5]. Joints in adults become affected more often than in children [6]. The most common localization is large joints of lower limbs; elbow and wrist joints are affected less often. In the patient, whose condition is described above, the inflammatory process involved ankle joints. Articular syndrome may be accompanied by myalgiae (muscle ache) and edemata of lower limbs [2]. Articular syndrome rarely lasts longer than 1 week.

Abdominal syndrome caused by gastrointestinal tract lesion is observed in approximately 2/3 of all patients [5]. It is characterized by spastic abdominal pain, nausea, emesis and gastrointestinal hemorrhage (moderately manifested non-life-threatening hemorrhages are observed often, in up to 50% of cases; severe hemorrhages are less common; life-threatening hemorrhages are observed in 5% of cases at most). Such complications as intestinal intussusception, perforation or peritonitis may occur [1]. Intussusception as a complication was identified in this patient by means of pneumography and unfolded at the inpatient hospital.

Endoscopic examination reveals hemorrhagic or erosive duodenitis, less often – gastric or intestinal erosions (of any localization, including rectum); they were also present in the patient's clinical pattern.

Renal syndrome rate varies from 10% to 60%, according to different data sources [3, 4]. It usually develops after occurrence of other signs of the disease, sometimes even 1-3 weeks after onset of the disease; however, in rare cases it may be the first symptom. Clinical signs of kidney damage may vary. It is usually characterized by isolated micro- or macroglobulinuria, which may sometimes be accompanied with moderate proteinuria. In most cases, these changes resolve without any sequelae; however, some patients develop glomerulonephritis [7]. Nephrotic syndrome may also develop.

Lungs and nervous system are only rarely affected. Patients may suffer from pulmonary bleeding and pulmonary hemorrhages [1, 3], as well as from encephalopathies and small changes of mental status [1, 3]. Severe headache, convulsions, cortical hemorrhages, subdural hematomata and even cerebrovascular accidents may occur. Polyneuropathy may develop [2, 7].

Scrotal lesions are observed in one of three children; they are manifested with scrotal edema due to hemorrhages in scrotal vessels [4].

Numerous clinical signs of hemorrhagic vasculitis described above have been observed in the patient many times.

Conclusion

In most cases, hemorrhagic vasculitis may be accompanied by intense abdominal pain syndrome. A surgeon responsible for follow-up of children with hemorrhagic vasculitis is required to pay particular attention to details of clinical course and be able to differentiate natural development of a complex of symptoms of hemorrhagic vasculitis from complicated course of the disease, which requires urgent surgical correction.

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Conflict of interest

The authors declared they have no competing interests to disclose.

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