CASE REPORT

Systemic lupus erythematosus (SLE): an unusual cause of ileocolic intussusception

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Accepted 30 October 2017

SUMMARY

Intussusception is a rare diagnosis in adults. Gastrointestinal involvement is not unusual in systemic lupus erythematosus (SLE).¹ A case of intussusception as first presenting symptom of SLE is described. A 50-year-old woman of Chinese origin came to the emergency room with symptoms of high fever, nausea, vomiting and weight loss of 10 kg. Three months before, an episode with abdominal pain due to ileal invagination in the colon occurred. Laboratory analysis showed: thrombocytopaenia, lymphopaenia, positive antinuclear antibody, antidouble-stranded DNA, Coombs tests and low C3 in combination with an active urine sediment and proteinuria. CT and positron emission tomography scan showed lymphadenopathy, but no other abnormalities. Axillar lymph node biopsy showed no abnormalities. The diagnosis new-onset SLE was made. Prednisone treatment was started and soon thereafter tapered, azathioprine and hydroxychloroquine were added. During follow-up, the SLE remained in remission. SLE is a rare cause of ileocolic intussusception and ileocolic intussusception may be the first presenting symptom.

BACKGROUND

Systemic lupus erythematosus (SLE) is a complex chronic inflammatory disease of unknown cause with a wide cascade of symptoms.² Abdominal pain does have an extensive differential diagnosis but gastrointestinal involvement is not unusual in SLE.^{3,4}

Many causes of abdominal pain are described in lupus patients, such as lupus peritonitis, severe lupus vasculitis presenting as mesenteric thrombosis or gastrointestinal perforation.^{5–9} In addition, the possible contributing factor of SLE in jejunal diverticulosis has been described.¹⁰

An intussusception as presenting symptom of SLE is rare; only six cases have been described in SLE. Four patients had an intussusception as initial presentation of SLE; in two of those patients, it was secondary to mesenteric vasculitis.^{4,9} Our case report describes a patient with an ileocolic intussusception as first presenting symptom of SLE.

CASE PRESENTATION

A 50-year-old woman of Chinese origin was admitted to our hospital with various complaints since 3 weeks: high fever, nausea, vomiting and, because of these complaints, weight loss of 6 kg in 3 months. Previous medical history was unremarkable besides constipation. Three months before this

presentation, she was seen at the emergency room for abdominal pain due to ileal invagination in the colon. An inflammation of Bauhin's valve and colitis were found at colonoscopy, biopsy revealed aspecific reactive changes possible due to the invagination, but an infectious or drug-induced cause (patient used ibuprofen) could not be excluded. After 2 months, another colonoscopy showed spontaneous regression of the abnormalities.

At this new disease episode, physical examination showed a cachectic woman (length: 1.64 m and weight: 46 kg vs 56 kg in 2012), with a temperature of 38.9°C, blood pressure of 115/65 mm Hg, heart rate of 93 beats per minute and respiration rate of 20 per minute. Notable was the white plaque on the dorsum of the tongue. There was diffuse abdominal tenderness, especially in the left lower quadrant.

INVESTIGATIONS

Laboratory analysis showed a pancytopenia with a normal C reactive protein level and a very high level of serum ferritin (24 550 µg/L). Urinary analysis revealed dysmorphic erythrocytes and an elevated protein excretion (microalbuminuria of 1180 mg/24 hours).

Immunological analysis showed a positive antinuclear antibody (ANA) and, antidouble-stranded DNA (117 UI/mL) and a decreased C3 level. Direct Coombs test was positive. Soluble-interleukin 2 (IL-2) receptor was 9200 (normal value <2500 pg/mL). Serological testing for anticardiolipin antibodies, lupus anticoagulants, rheumatoid factor, anti-cyclic citrullinated peptide (CCP), antineutrophil cytoplasmic antibodies (ANCA), anti-SSA/Ro, anti-SSB/La and anti-Smith was negative.

There were no arguments for an infectious cause because in several serological tests, such as HIV, Epstein-Barr virus, cytomegalovirus, parvovirus, hepatitis, tuberculosis, *Coxiella burnetii* were negative. A CT scan of chest and abdomen showed lymphadenopathy of numerous small lymph nodes in axilla, para-iliacal, mediastinal and hilar. Positron emission tomography scan confirmed the lymphadenopathy, but no other abnormalities were found. Upper gastrointestinal endoscopy showed a *Candida* oesophagitis.

Bone marrow examination revealed slight abnormalities which could be either reactive or a beginning myelodysplastic syndrome. Hemophagocytosis was not found. Axillar lymph node biopsy showed no abnormalities, but it should be noted that our patient at this moment already was treated

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To cite: Glijn N, Korswagen L-A, Lam-Tse W-K. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220185

with corticosteroids and the resected lymph node was only 2 mm small so this was not very representative.

DIFFERENTIAL DIAGNOSIS

The diagnosis of SLE with secondary macrophage activation syndrome/hemophagocytic lymphohistiocytosis (HLH) was made. For SLE, this was based on thrombocytopenia, lymphopenia, positive ANA, positive antidouble-stranded DNA, positive Coombs test, low C3 and the abnormalities in the urine sediment and proteinuria.

According to the haematologist, a haematologic malignancy, such as lymphoma, was unlikely, mainly because the lymph nodes were very small. Furthermore, 3 months earlier, there were no haematologic abnormalities; therefore, the rapid onset of pancytopenia was more indicative of an infection or autoimmune disease.

There is an overlap between criteria for SLE and HLH. Arguments for HLH are fever, pancytopenia, elevated ferritin level, elevated soluble IL-2 receptor and hypertriglyceridaemia.¹¹ Other criteria such as hemophagocytosis in bone marrow or splenomegaly were not present. Testing for natural killer cell activity has not been performed. HLH could be secondary to many diseases, but in this case, the most likely cause was a new-onset SLE.^{12 13}

TREATMENT

HLH is an aggressive and life-threatening syndrome, which generally needs aggressive immune suppression, for example, a combination of at least corticosteroids, ciclosporin and/or etoposide.¹³ However, if HLH occurs in association with an underlying triggering condition, in this case SLE, treatment of this condition alone in a not acutely ill patient could be sufficient.¹¹ The patient was treated with prednisone at a dose of 50 mg once daily with good and quick clinical response. Azathioprine was started as steroid-sparing agent and hydroxychloroquine was added.

OUTCOME AND FOLLOW-UP

The body temperature normalised, the pancytopenia resolved and the urinary sediment normalised. After 1 month, the patient was discharged. For the *Candida* oesophagitis, she was treated with fluconazole for 14 days. Prednisone was tapered. During follow-up, the SLE remained in remission.

DISCUSSION

SLE is a rare disease of unknown cause in which deposition of pathogenic autoantibodies and immune complexes can cause a broad variety of symptoms.¹⁴ It can affect almost every organ system and could be a life-threatening disease.

Abdominal pain in patients with SLE is not uncommon. It can be due to more common problems like (lupus) peritonitis, peptic ulceration, acute pancreatitis and cholecystitis, but also more rare causes like hepatitis, lupus mesenteric vasculitis, splenic artery aneurysm and jejunal diverticulosis have been described.^{5–10}

Intussusception is a condition in which a proximal segment of the gastrointestinal tract folds into the lumen of the adjacent segment.³ Although it is mainly seen in childhood, it is a rare condition in adults (5% of all cases of intussusception).³

In the small intestine, an intussusception can be secondary either to the presence of intraluminal or extraluminal lesions (inflammatory lesions, Meckel's diverticulum, postoperative adhesions, lipoma, lymphoma and metastases), but intussusception in the large bowel is more likely to be caused by a malignant aetiology.³ Usually the CT scan can reveal the invagination and

the potential cause. The appearance of a bowel-within-bowel configuration with or without contained fat and mesenteric vessels is pathognomonic for intussusception.⁵

The pathogenesis of the intussusception in our case might be related to the SLE and could very well be the initial presentation of the disease. Only six cases of intussusception in SLE patients have been previously reported in the literature. In three of them, it was secondary to lupus mesenteric vasculitis (LMV), in one lymphadenopathy was thought to be the leading cause, one was secondary to altered peristalsis of oedematous intestine and one was related to Burkitt's lymphoma (BL).^{4–9}

Hermann^{3 4} published the first case report about a 5-year-old child with already known SLE who had an intussusception secondary to LMV. The mechanism of LMV causing intussusception is not well understood.⁴ A possible explanation is that vascular necrosis is caused by diffuse vasculitis with partial devitalisation of a bowel segment. This can lead to interruption of the normal neuromuscular function, with concomitant intussusception and risk of venous infarction requiring intestinal resection.⁴ This is an extremely serious complication with a high mortality.¹⁵

Abdominal CT imaging has an important role in the early detection of LMV and provides accurate imaging of lesions.⁵ In our case, the CT scan did not show pathognomonic findings for LMV such as: dilated bowel, bowel wall thickening, comb sign or ascites. The spontaneous remission of the intussusception, without immunosuppressive treatment, is another argument against LMV as underlying cause.

Intussusception caused by BL is rare, often with misleading symptoms and difficult to diagnose. Two cases of BL-related ileocecal intussusception were operated by Yagmur *et al.* They pointed out that complete resection is required and improves patient survival.¹⁵

Another case report described a lupus patient with fungal infection with intussusception. It is unclear whether there is an association of these conditions.⁶ In our patient, intussusception took place months before the *Candida* oesophagitis was found, but this latter could have been present; therefore, it could play a role in the pathogenesis of intussusception in this case.

The intussusception took place 3 months before this presentation with SLE with critical illness and macrophage activating syndrome. We find it not likely that HLH plays a main role in the development of the intussusception, since this syndrome develops in a short period of time.

Like mesenteric vasculitis, HLH is a sign of highly active lupus disease and, in patients with intussusception and suspicion of SLE, physicians should in general be aware of possible other severe complications of lupus.

Also, we have to be aware of the possibility of another factor contributing to the intussusception. Her poor nutritional status could have predisposed this lupus patient to develop an intussusception.

However, in the absence of arguments for malignancy, tuberculosis or other systemic inflammatory disease, we find it highly

Learning points

- ▶ Be aware of underlying pathology in adult patients with an intussusception.
- ▶ Systemic lupus erythematosus (SLE) is a rare cause of ileocolic intussusception.
- ▶ Ileocolic intussusception can be the first presenting symptom of SLE.

likely that the intussusception could be the first presenting symptom of SLE.

In conclusion, a 50-year-old patient presented with an ileocolic intussusception, 3 months before she was diagnosed with SLE. There was no evidence for mesenteric vasculitis nor signs of other pathology like malignancy or an infectious cause. Therefore, it is likely that the intussusception was the initial presentation of SLE in this patient.

Contributors NG participated in collecting the patient information and writing of the case report. L-AK participated in collecting the patient information and writing of the case report. W-KL-T initiated the case report and participated in writing of the case report.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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