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# Aseptic abscess syndrome as first manifestation of Crohn's disease – a case report

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## Abstract

**Background** There exist multiple extraintestinal manifestations of inflammatory bowel disease. The most common are arthritis, aphthous stomatitis, or uveitis. Aseptic abscess syndrome is not usually included among these extraintestinal manifestations. In our case report, we present a rare case of aseptic abscess syndrome as the first manifestation of inflammatory bowel disease.

**Case presentation** We present the case of a 10-year-old girl whose only initial medical issue was recurrent submandibular lymphadenitis unresponsive to standard antibiotic therapy. A broad differential diagnosis was initiated to exclude an infectious etiology. Eventually, it was necessary to proceed with extirpation of the suspected lymph node. Histological examination showed suppurative granulomatous inflammation, so it was further examined for noninfectious cause. Blood tests revealed positivity of ASCA antibodies (Anti-*Saccharomyces cerevisiae*) in both IgA and IgG classes. Despite absence of typical gastrointestinal symptoms, bowel ultrasound was performed, followed by magnetic resonance enterography. Both showed inflammatory changes in the terminal ileum. Subsequent endoscopy of the gastrointestinal tract and histological examination of biopsy specimens confirmed a diagnosis of Crohn's disease with terminal ileum and rectum involvement. A standard treatment based on current guidelines led to remission without recurrence of lymphadenitis.

**Conclusions** In cases of lymphadenitis that does not respond to standard antibiotic treatment, diagnosis of aseptic abscess syndrome should be considered as a potential etiology and, subsequently, inflammatory bowel disease should be investigated, given that this syndrome is associated with inflammatory bowel disease in as many as 70% of cases. To our knowledge, this is the first published case report describing aseptic abscess syndrome affecting cervical lymph nodes as an extraintestinal manifestation of pediatric Crohn's disease.

**Keywords** Aseptic abscess syndrome, Extraintestinal manifestation, Inflammatory bowel disease, Crohn's disease, Pediatrics

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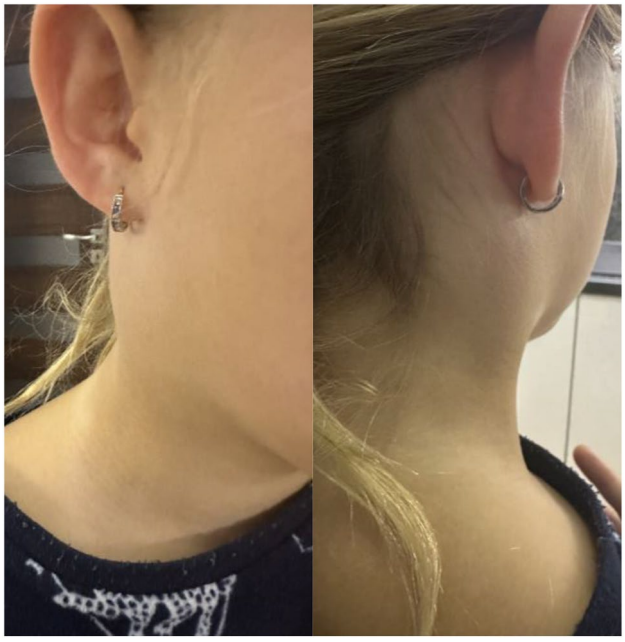

**Fig. 1** Appearance of right-sided cervical lymphadenitis

## Background

Inflammatory bowel diseases (IBDs) are chronic conditions affecting the gastrointestinal tract (GIT) and can be classified into three subtypes: Crohn's disease (CD), ulcerative colitis (UC) and unclassified inflammatory bowel disease (IBD-U). Clinical presentation of IBD is variable, but abdominal pain, diarrhea (in some cases with blood), and weight loss are typical signs of GIT affection. Extraintestinal manifestation (EIM) – affection of extraintestinal organs – is present in approximately 17–28% of pediatric patients [1–3]. EIMs are more common in CD (22.5%) than UC (10.3%) [1] and most often involve the joints, skin, oral mucosa, or eyes [4, 5]. In this case report, we highlight a rare symptom – aseptic abscess syndrome – as the first manifestation of Crohn's disease.

## Case report

We present the case of a 10-year-old girl from a high-risk twin pregnancy after in vitro fertilization, born at 32 weeks of gestation by caesarean section, with uncomplicated postnatal adaptation. There was no significant family history. The patient was followed up in allergology for pollen and eggplant allergy and recurrent wheezing bronchitis and also by an ophthalmologist for *sicca* syndrome.

The patient was scheduled for endonasal removal of a bleeding polyp of the nasal septum. Simultaneously at admission, the patient presented with right-sided cervical lymphadenitis (Fig. 1). Despite successful endonasal surgery the lymphadenitis persisted. On the ultrasonography (US) there was unilateral lymph node (LN) enlargement (33 × 19 mm). Serum inflammatory parameters were low. Infectious etiology of lymphadenitis was not confirmed. Odontogenic origin was also ruled out. Empirical antibiotic treatment (amoxicillin–clavulanate) was not effective. On the contrary, after 1 month of antibiotic treatment the size of the observed LN had progressed (42 × 22 mm). Therefore, extirpation was indicated. The material was sent for histological examination, which showed granulomatous suppurative lymphadenitis (Fig. 2). After extirpation, the patient was without medication and in good condition.

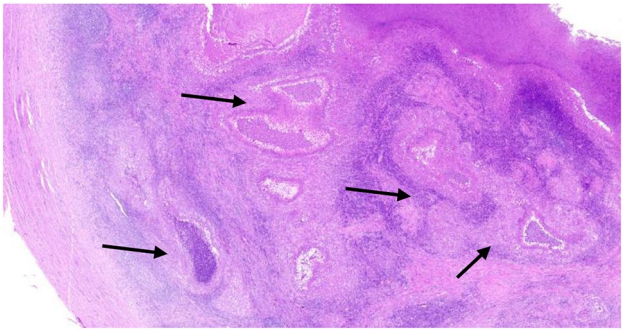

**Fig. 2** Histological appearance of extirpated lymph node. Arrows indicate large suppurative granulomas

Seven months after extirpation, the patient presented again with enlarged LN submandibularly on the left, 27 × 15 mm according to US. Serological examination of infectious agents was again negative. Antibiotic treatment (clarithromycin) did not result in the condition's improvement. On the contrary, there was again progression of LN size on US and the patient reported severe pain. Torticollis and edema of surrounding tissues were observed. The patient was admitted to the Department of Pediatric Infectious Diseases for further investigation. She had no fever during the whole course of the disease. Due to persistence of lymphadenitis despite combined parenteral antibiotic treatment (clindamycin + gentamicin) and suppuration on cervical CT scan, extirpation of suspected LN was indicated. Histology confirmed granulomatous suppurative lymphadenitis. PCR examination of LN material excluded infectious etiology. Detailed and broad examinations to clarify the etiology were performed (see Table 1). After an uncomplicated postoperative period, the patient was discharged in good condition for further outpatient care.

After discharge, the patient newly developed daily abdominal pain, localized periumbilically, with relief after defecation. The patient's stool passed once a day and was without pathological admixture. In retrospect, an unexplained weight loss was reported (6 kg over the past 6 months). After excluding infectious diseases of the GIT and considering the ASCA positivity, we followed up with bowel US and subsequently magnetic resonance enterography. These showed inflammatory changes in 5–10 cm

**Table 1** Examinations performed to identify the etiology of lymphadenitis

| | | |
|---|---|---|
| Laboratory tests | CRP level | Normal |
| | Biochemistry | Negative |
| | Sedimentation rate | 62/102 mm/hour |
| | Blood count | Without abnormalities |
| | Serology for felinosis, toxoplasmosis, tularemia | Negative |
| | Autoantibodies | ANA positive (1:800)<br>ssDNA negative<br>dsDNA negative<br>ENA negative<br>ASCA IgG positive (60.6 U/ml), ASCA IgA positive (> 100 U/ml)<br>ANCA negative<br>tTGA IgA negative |
| | Cell-mediated (including burst test), humoral immunity | Without abnormalities |
| | Stool culture for Campylobacter jejuni, Yersinia enterocolica | Negative |
| Imaging methods | Cervical CT scan | Bilateral lymphadenitis, signs of suppuration in the left largest LN |
| | Chest X-ray | Negative |
| LN extirpation | Histology | Granulomatous suppurative lymphadenitis |
| | Flow cytometry | No clonal population |
| | Acid-fast stain | Negative for acid-fast rods |
| | Grocott methenamine silver stain | Negative |
| | Cultivation | Negative |
| | PCR *Bartonella* spp., *Borrelia burgdorferi*, *Brucella* spp., *Francisella tularensis*, *Mycobacterium tuberculosis*, *Mycobacterium* spp., *Toxoplasma gondii*, *Tropheryma whipplei* | Negative |
| Rheumatology | Sarcoidosis | Excluded |
| | Sjogren's syndrome | Excluded |
| Other | Stomatology examination | Odontogenic origin of infection excluded |

CRP – C reactive protein, ANA – antinuclear antibody, ssDNA – single-stranded DNA, dsDNA – double-stranded DNA, ENA – extractable nuclear antigen, ASCA – anti-*Saccharomyces cerevisiae*, ANCA – anti-neutrophil cytoplasmic antibody, tTGA – tissue transglutaminase antibody

of terminal ileum and mild inflammatory changes of the appendix (Fig. 3). With respect to this information, we tested stool for fecal calprotectin, which was surprisingly low (40.2 ug/g), and fecal occult bleeding, which was positive (182 ng/ml).

These findings led us to suspicion of IBD, so the patient was indicated for endoscopic examination of the GIT. Macroscopically on esophagogastroduodenoscopy there were multiple aphthous lesions in the gastric antrum. However, these had no histological correlation. Subsequent colonoscopy showed mucosal edema and cobblestone appearance in the initial 5 cm of the terminal ileum, followed by two aphthous lesions approximately 10 cm apart (Fig. 4); a few additional aphthous lesions were found in the rectum; the rest of the examined bowel was normal. Histological examination showed chronic granulomatous inflammation of the terminal ileum, cecum and colon ascendens (Fig. 5). A diagnosis of Crohn's disease was established according to the revised Porto criteria [6]. Remission was induced using a Crohn's disease exclusion diet (CD-ED) and maintenance treatment by azathioprine. The established therapy led to long-term remission of the disease. Furthermore, the patient was without recurrence of lymphadenitis and adverse effects of treatment and is regularly followed up by her pediatric gastroenterologist.

## Discussion

Extraintestinal manifestations of IBDs encompass a wide range of symptoms and can be defined as inflammatory processes outside the gut but directly related to the existence of IBD [7]. According to the European Crohn's and Colitis Organisation, these can be classified as classical EIM, systemic consequences of IBD, and conditions loosely associated with IBD [8]. Strict classification of EIM, however, remains a matter of debate [8]. At least one of the EIMs affect between 17% and 28% [1–3] of children with IBD. EIMs present more frequently in patients with CD (22.5%) than with UC (10.3%) [1]. According to Heyman et al. [2], EIMs precede the diagnosis of IBD in childhood in 6% of cases, with an incidence of 9% after 1 year of IBD, 19% after 5 years, and even 29% after 15 years. Another study has reported a much higher incidence of EIM (up to 27.6% of cases) at the time of IBD diagnosis [1].

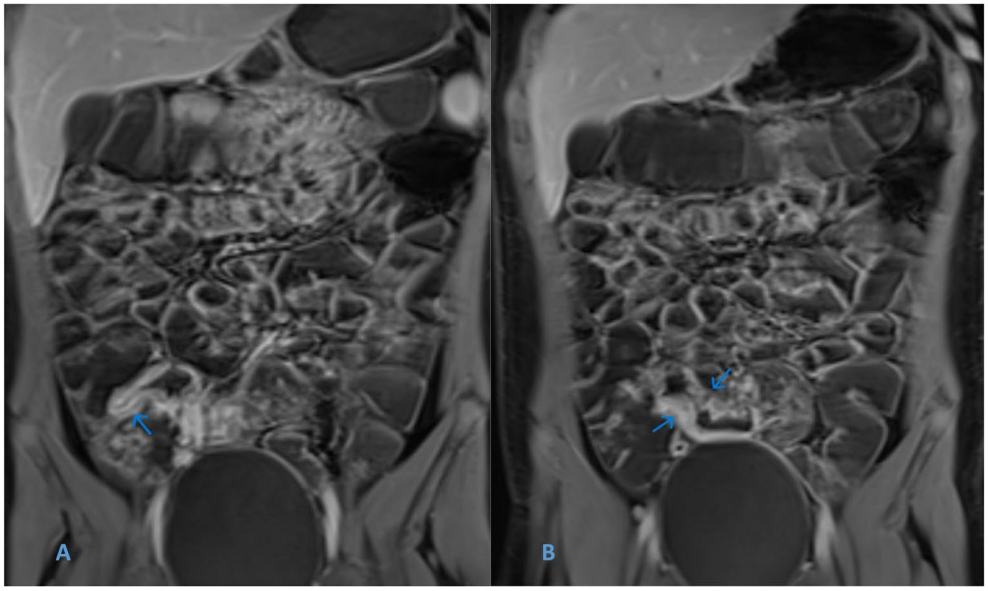


**Fig. 3** Magnetic resonance enterography. Arrows indicate inflammatory changes of ileocecal valve (**A**) and ansa ultima of terminal ileum (**B**)

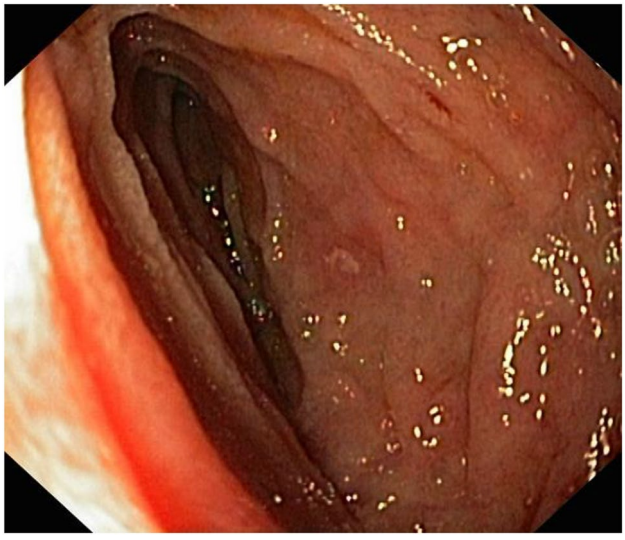

**Fig. 4** Appearance of terminal ileum on coloscopy

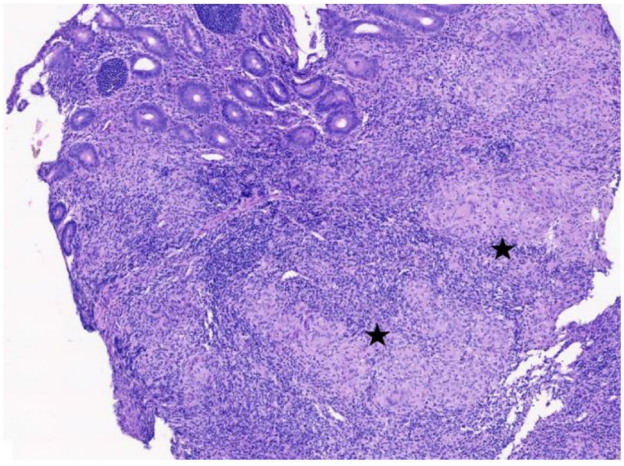

**Fig. 5** Histological appearance of terminal ileum. Stars indicate non-necrotizing granulomas

Although EIMs most commonly affect the skin and oral mucosa, joints, bile ducts, and eyes [4, 5], as presented in Table 2, almost any organ can be affected [5].

The most common EIMs are arthritis (8–26%) and aphthous stomatitis (7–21%) [1, 2]. These two EIMs typically precede the diagnosis of IBD. On the contrary, osteopenia or osteoporosis are observed after the diagnosis of IBD [1, 2]. Other common EIMs are uveitis and erythema nodosum [1].

Rare EIMs include metastatic Crohn's disease, orofacial granulomatosis, and pyostomatitis vegetans. Metastatic Crohn's disease is a granulomatous dermatitis characterized by the presence of non-necrotizing granulomas identical to those found in CD in the GIT [9, 13]. The skin lesions resemble reddish, ulcerated nodules localized to the skin folds, under the breasts [13, 14], and/or on the vulva or penis [13–15]. Orofacial granulomatosis presents as swelling of the lips and face and also is characterized by the presence of non-necrotizing granulomas [9]. Pyostomatitis vegetans represents a more severe form of aphthous stomatitis – it manifests as multiple erosions and pustules resembling cobblestones in the oral cavity [5, 9]. It is more common in patients with UC. In patients with CD, it is associated with zinc deficiency [5, 9].

Granulomatous lymphadenitis is most often of infectious origin (e.g., in cases of tularemia, felinosis, toxoplasmosis, or mycobacteriosis). Less often it is noninfectious (e.g., in sarcoidosis; see Table 3) [16]. In his review, Asano described granulomatous lymphadenitis in CD as the

**Table 2** Extraintestinal manifestations and systemic complications of IBD. Adapted from [8–12]

| | Extraintestinal manifestations | Systemic complications of IBD |
|---|---|---|
| Skin and mucosae | Erythema nodosum | |
| | Pyoderma gangrenosum | |
| | Aphthous stomatitis | |
| | Angular stomatitis | |
| | Pyostomatitis vegetans | |
| | Orofacial granulomatosis | |
| | Sweet syndrome | |
| | Metastatic Crohn's Disease | |
| Joints | Peripheral arthritis | Osteopenia, osteoporosis |
| | Axial arthritis | |
| Hepatobiliary tract | Primary sclerosing cholangitis | |
| | Autoimmune hepatitis | |
| Pancreas | Pancreatitis | |
| Eye | Episcleritis | |
| | Scleritis | |
| | Uveitis | |
| Hematological | | Anemia |
| | | Venous thrombosis |
| Other | Aseptic abscess syndrome | Growth retardation |
| | | Delayed puberty |

**Table 3** Etiology of granulomatous lymphadenitis. Adapted from [16]

| Infectious etiology | Noninfectious etiology | |
|---|---|---|
| Tularemia | Sarcoidosis | |
| Felinosis | Berylliosis | |
| Tuberculosis | Immunodeficiency | Primary biliary cirrhosis |
| Atypical mycobacteriosis | | Sjogren's syndrome |
| Toxoplasmosis | | Crohn's disease |

noninfectious cause of granulomatous lymphadenitis [16]. Practically speaking, scattered granulomas without necrotic changes or suppuration (sarcoid-like) can be found in mesenteric lymph nodes in patients with CD [17].

We believe that this case represents aseptic abscess syndrome (AAS), a rare disorder associated with systemic inflammatory conditions [18–21], which is characterized by a sterile collection of neutrophils in a variety of organs, typically liver or spleen, rarely lymph node. Histologically, these aseptic abscesses are created by central collection of pus surrounded by granulomatous reaction [18–20]. This syndrome affects patients with systemic inflammatory conditions and, according to data available in the literature, it is associated with IBD in as many as 70% of cases [19]. Typical and similar to our case is also the fact that patients with AAS are often misdiagnosed and initially are treated unsuccessfully for bacterial infection [19]. Most patients with AAS respond well to immunosuppression and achieve remission [18, 19].

We conclude from our patient's story that AAS should be included among the rare EIMs of IBD. We base this opinion on the fact that, despite extensive diagnostic work-up, no other objective cause of granulomatous suppurative lymphadenitis was identified and the therapy undertaken led to remission without further recurrence of lymphadenopathy. This is supported by Bollega et al. who report AAS as a rare EIM and described two cases of AAS in IBD patients [21]. Our case also meets the set of common characteristics of AAS that André et al. used to identify AAS in their study [18]. Indeed, in a case when AAS is suspected endoscopic examination of the GIT should be considered, as AAS could be the first presentation of IBD.

## Conclusion

IBDs typically manifest with abdominal pain, unformed stools (often with blood), and weight loss. As many as one-quarter of pediatric patients present with extraintestinal symptoms. Arthritis, aphthous stomatitis, or uveitis are the most common, but almost any organ can be affected. Our case report demonstrates that recurrent lymphadenitis unresponsive to standard treatment should be suspected as AAS, which may be a rare and first manifestation of IBD.

**Abbreviations**

| | |
|---|---|
| AAS | Aseptic abscess syndrome |
| CD | Crohn's disease |
| CD-ED | Crohn's disease exclusion diet |
| EIM | Extraintestinal manifestation |
| GIT | Gastrointestinal tract |
| IBD | Inflammatory bowel disease |
| LN | Lymph node |
| UC | Ulcerative colitis |
| US | Ultrasound |

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**Author contributions**
MA and MH conceptualized and drafted the manuscript. MA searched the literature. PJ revised and supervised the manuscript. MU supervised the otorhinolaryngological care of the patient. MJ performed histological examination of the specimens. All authors have read and agreed to the published version of the manuscript.

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**Data availability**
No datasets were generated or analysed during the current study.

## Declarations

**Ethics approval and consent to participate**
Not applicable.

**Consent for publication**
Written informed consent was obtained from the parent of the patient.

**Competing interests**
The authors declare no competing interests.

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## Publisher's note