Case Report

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Navigating the complexity of Churg-Strauss syndrome presenting as acute abdomen: a comprehensive review and case report

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Introduction and importance: Churg-Strauss syndrome (CSS) is a rare multisystemic condition characterized by asthma, blood and tissue eosinophilia, and vasculitis. The purpose of this work is to present a detailed overview of CSS, focusing on its epidemiology, clinical symptoms, histological criteria, gastrointestinal involvement, and therapy.

Case presentation: The authors report a case of a 40-year-old woman with CSS who had peripheral eosinophilia, small vessel vasculitis, and bronchial asthma. Diagnosed with symmetric polyarthritis six months ago, experienced abdominal pain, vomiting, and loose faeces. Diagnostic tests revealed CSS with systemic involvement. In addition, we undertake a literature analysis to emphasize essential elements of CSS, such as its rarity and the difficulties in diagnosing and managing it.

Clinical discussion: CSS can cause gastrointestinal symptoms including stomach pain, diarrhoea, mucosal ulcers, rectal bleeding, and bowel perforations. Corticosteroids and immunosuppressives are routinely used in treatment, with caution due to the risks of long-term steroid use. The goal of treatment should be to induce remission while minimizing side effects.

Conclusion: CSS is a rare condition, with an annual incidence of 2.4 per million and a prevalence of 1.3 per 100 000. The illness typically presents with necrotizing vasculitis, extravascular granulomas, and eosinophilic tissue infiltration. CSS is a complex and rare condition that requires high clinical suspicion, especially when patients present with gastrointestinal symptoms in addition to asthma and eosinophilia. This case study adds to our understanding of CSS and emphasizes the significance of a holistic strategy for its management.

Keywords: churg-strauss syndrome, corticosteroids, eosinophilic granulomatosis with polyangiitis, gastrointestinal involvement, vasculitis

Introduction

Churg-Strauss syndrome (CSS), first identified by J. Churg and L. Strauss, is a disorder that is similar to polyarteritis nodosa (PAN) but separate from it. Asthma, blood and tissue eosinophilia, and small vessel vasculitis are the hallmarks of the multisystem illness eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as CSS[1]. According to a study done by R. A. Watts *et al.*[2] between 1988 and 1994 on 414,000 people in a stable, ethnically homogeneous community, the annual incidence of CSS was estimated to be 2.4 per million. Another retrospective study performed by G. Haugeberg *et al.*[3] showed a prevalence of 1.3 per 100 000.

HIGHLIGHTS

- Churg-Strauss syndrome (CSS) is a rare multisystem syndrome with complex symptoms.
- Gastrointestinal involvement: CSS manifests as stomach pain, ulcers, and bowel perforations.
- Epidemiology insights: CSS's rarity with incidence at 2.4 per million.
- Histological hallmarks: Necrotizing vasculitis, eosinophilic infiltration, and granulomas.
- Holistic CSS management: Balancing corticosteroids and alternative therapies.

The three key histological criteria of this condition are necrotizing vasculitis, eosinophilic tissue infiltration, and extravascular granulomas, which are rarely present together[4]. The condition is peculiar given that it combines asthmatic symptoms with hypereosinophilic diseases and ANCA-associated vasculitis (AAV). As a result, a complete understanding of its pathogenesis is still unclear[5].

Gastrointestinal tract (GIT) involvement and patient presentation in CSS can range from abdominal pain, diarrhoea, and nausea to mucosal ulcers (notably in the duodenum), rectal bleeding, ischaemic bowel, and bowel perforation needing

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laparotomy[6]. In 96 patients with EGPA treated between 1963 and 1995, Guellivin and colleagues did a retrospective study and found that 30% of patients experienced GI symptoms such as melena, hematemesis, or stomach discomfort, which indicated intestinal vascular involvement. They also discovered that a negative outcome is associated with intensive GIT involvement[7].

We describe a 40-year-old woman who had peripheral eosinophilia, small vessel vasculitis, and bronchial asthma suggestive of CSS. The patient improved after receiving low-dose steroids and cyclophosphamide pulse therapy during follow-up.

This case report has been reported in line with the SCARE Criteria[8].

Patient information

A 40-year-old woman presented to the emergency department with acute abdomen, vomiting, and loose faeces for 3 days. Her medical history revealed that she has hypertension and asthma. Six months ago, the patient was diagnosed with symmetric polyarthritis; afterwards, she had a right foot drop and paraesthesia in both her lower and upper limbs, and further testing for suspected vasculitic neuropathy came back negative. She was prescribed steroids for her symptoms. The patient used steroids for a month before stopping on her own. Subsequently, she experienced polyarthralgia, a rash, and limb weakness in both the upper and lower extremities.

Clinical findings

On examination, the patient had a desquamating, palpable purpuric rash throughout both lower limbs as well as right hypochondriac pain. Upon neurological examination, there was distal muscle atrophy in both the upper and lower limbs, decreased grip strength on both sides of the hands, right foot drop, absence of all deep tendon reflexes, and distal sensory impairment in both the upper and lower limbs without any cranial nerve involvement.

Diagnostic assessment and interpretation

Investigation

Investigations revealed a decreased serum C3, a normal C4, and an elevated white cell count of 42 300/mm^3, with 10% eosinophils and a shift to the left.

Radiology

MRCP revealed cholelithiasis with sludge in the gallbladder and a dilation of the common bile duct (CBD) with sludge or tiny calculi. Specific symptoms and pneumoperitoneum detected by abdominal ultrasonography were suggestive of hollow viscus perforation.

Diagnosis

Because bronchial asthma, small vessel vasculitis, and peripheral eosinophilia were present, a definitive diagnosis of CSS with systemic involvement was made.

Intervention

Surgical intervention—exploratory laparotomy

A solitary ileal perforation measuring 5 mm in size, as seen in Fig. 1, was discovered during a laparotomy on the patient, along with a region of potential perforation 15 cm from the ileo-caecal (IC) junction and numerous ulcers throughout the ileum. The perforation was sealed, and there were no complications during the healing process. HPE from the edge of the perforation showed vascular-rich fibro-collagenous tissue with neutrophilic vasculitis.

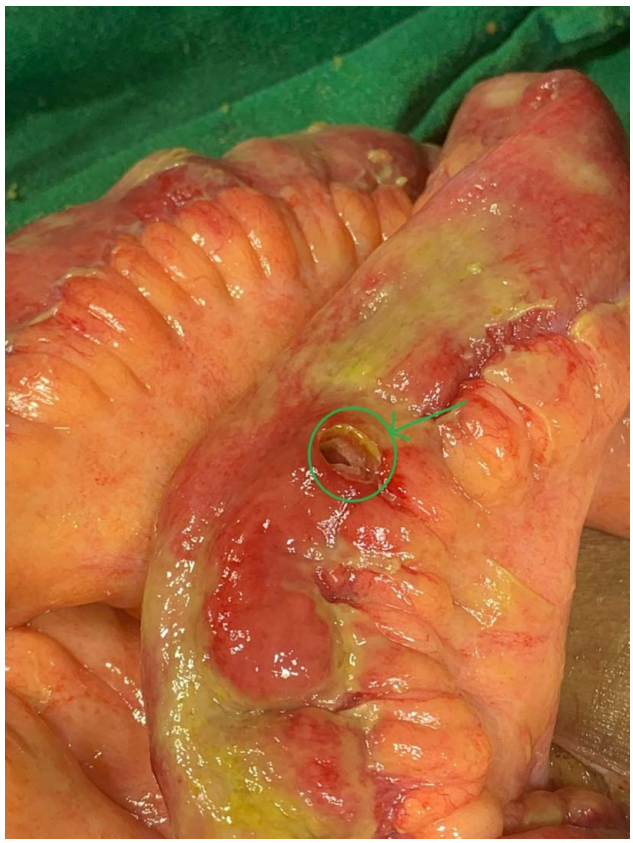

Figure 1. Intestinal perforation seen on exploratory laparotomy.

Follow-up and outcomes

Postoperative workup

Postoperative workup revealed a white cell count of 12 000/mm^3, with a differential count showing 48% eosinophils; p-ANCA and c-ANCA were negative; an elevated RA factor of 494 U/L; anti-CCP was negative; and a nerve conduction study showed severe sensory motor axonal neuropathy.

Medical management

During the subsequent examination, the patient received cyclophosphamide pulse therapy and low-dose steroids, resulting in significant improvement. The patient's follow-up was inconsistent and led to missing data.

Discussion

CSS, also known as EGPA, is a condition characterized by inflammation of small blood vessels, an increase in eosinophils in the tissues and blood, and the presence of asthma. It is associated with antineutrophil cytoplasmic antibodies (ANCA) and is

believed to be caused by T-helper (Th) 2 cells and their secretion of cytokines[9,10].

One of the complications of EGPA is gastrointestinal involvement, which can lead to symptoms such as nausea, vomiting, diarrhoea, ulcers in the duodenum, rectal bleeding, reduced blood flow to the intestines, and in severe cases, intestinal perforation. Studies have shown an increasing rate of gastrointestinal complications in patients with EGPA[11,12].

Intestinal perforation has been documented in cases of EGPA, often following the initiation of high-dose glucocorticoid treatment. The resected tissues in these cases showed signs of intestinal perforation, inflammation, oedema, bleeding, and vasculitis. Continuous administration of glucocorticoids has also been associated with EGPA vasculopathy and intestinal perforation[13,14].

The severity and prognosis of EGPA can be evaluated using the five-factor score, which takes into account organ involvement, clinical presentation, and biochemical markers. Patients with a score of 0, indicating no prognostic risk factors, can achieve remission with low-dose corticosteroids alone. However, around one-third of these patients may experience a recurrence of symptoms and require immunosuppressant medications[14].

EGPA can be categorized into two distinct disease phenotypes based on ANCA levels: ANCA positive, characterized by vasculitis symptoms, and ANCA negative, characterized by eosinophilic characteristics[5]. The disease is marked by hypereosinophilia and can cause organ damage through eosinophil infiltration and inflammation. It is also associated with elevated levels of blood IgG4 and IgE[15,16].

The progression of EGPA typically takes patients 3–9 years, starting with prodromal allergy symptoms, followed by an increase in eosinophils and tissue infiltration, and finally systemic signs of vasculitis[17]. Gastrointestinal involvement in EGPA can lead to various adverse physical symptoms, including ulcers, bleeding, and bowel perforation[6].

Treatment for EGPA often involves the use of corticosteroids, but long-term use can increase the risk of peptic ulcers and gastrointestinal bleeding[18]. To prevent these complications, alternative immunosuppressive drugs such as cyclophosphamide, azathioprine, methotrexate, and mycophenolate mofetil may be used. These drugs have shown efficacy in improving clinical outcomes and reducing the need for prolonged steroid use in other autoimmune diseases.

Limitations

The limitations of this case study on CSS are its reliance on a single incidence, the absence of regular patient follow-up, and a gap in the available data. The condition exhibits a wide range of variations, and the article examines a digestive symptom that may fail to take into account clinical distinctions. The prognosis of treatment is uncertain, and identifying CSS is difficult. Factors like as ethnic and geographical characteristics, publishing bias, and informed consent have a complex influence on patient opinions, making the issue more intricate. The scope of the case report is confined to medical knowledge up until January 2022, and any potential future developments may not be taken into account.

Conclusion

This case study focuses on the difficulties and complexities related to EGPA CSS) and its impact on the gastrointestinal system. It highlights the importance of being highly suspicious in persons who have an acute abdomen, blood eosinophilia, and a history of asthma. It is important to carefully evaluate treatment choices, such as corticosteroids and immunosuppressants, in order to achieve disease remission while minimizing any negative consequences. It is essential to create a detailed and targeted management strategy for the different presentations of EGPA, especially those affecting the gastrointestinal tract.

Ethical approval

There is no requirement for ethical consent.

Informed consent

The patient's consent is obtained after she has been informed about the study.

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Author contributions

Conceptualization: S.B., V.N.B., E.N.D., P.D., R.G.; Methodology: E.N.D., P.D., R.G.; Validation: P.D., R.G.; Formal analysis, Investigation: P.D., R.G.; Data curation: P.D., R.G.; Writing—original draft: S.B., V.N.B., E.N.D., P.D., R.G.; Visualization: S.B., V.N.B., E.N.D.; Writing—review and editing: P.D., R.G.; Editing: P.D.; Supervision, project administration: R.G.

Conflicts of interest disclosure

Not applicable.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

Dr Subbaraidu is the principal investigator and guarantor for the paper.

Data availability statement

The authors confirm that the data supporting the findings of this study are available within the article and its supplementary materials.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Patient Perspective

The patient tolerated the treatment well; there were no complaints of adverse reactions to pulse therapy.

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