Eosinophilic granulomatous vasculitis mimicking a gastric neoplasm

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Sir: A gastrointestinal presentation in eosinophilic granulomatous vasculitis syndromes (EGV) is rare. We describe a patient with an EGV, considered to be an atypical Churg–Strauss syndrome (CSS), presenting as a gastric neoplasm. In CSS gastrointestinal symptoms such as abdominal pain/diarrhoea or gastrointestinal bleeding occur in 44–89% of patients and pathological involvement of the gastrointestinal tract occurs in 33–92%.[1][2][3][4] Mortality attributable to gastrointestinal involvement is about 8%, and this is mainly from gastrointestinal bleeding or bowel perforation.[4] In spite of this, presentation with gastrointestinal symptoms is rare in EGV[3, 4] and we could find only one report in the literature of a patient with CSS who presented with diarrhoea and gastrointestinal bleeding three years before developing asthma.[3]

A 42-year-old woman presented with a 3-month history of severe loss of appetite, marked loss of weight and frequent vomiting with several episodes of haematemesis and malaena. She also complained of generalized weakness, and back pain. She had no dysphagia and her bowel habit was normal. She appeared emaciated and pale, and had a palpable irregular, 40 × 50 mm hard, mobile mass in the left hypochondrium. Her haemoglobin was 5.4 g/dl, with hypochromic microcytic red cells. The white cell count was 12.4×10^9/1 with 80% neutrophils, 15% lymphocytes, 5% eosinophils, and erythrocyte sedimentation rate was 130 mm in the first hour. A chest X-ray showed diffuse shadowing in the middle and lower zones of the right lung. Endoscopy showed an extensive nodular infiltrating plaque like lesion extending from the body of the stomach to the antrum partially obstructing the pylorus. Mucosal biopsies were normal.

On the fourth day after admission the patient developed a cough with haemoptysis, but she did not have dyspnoea or wheezing. Her clinical condition deteriorated. Palliative gastric bypass surgery was undertaken to reduce vomiting. Laparotomy revealed a mass in the distal half of the stomach, with relatively normal looking overlying mucosa suggestive of an infiltrative gastric neoplasm, and free fluid in the abdominal cavity. Full thickness biopsies from the ‘tumour’ were taken and a gastrojejunostomy performed. She made a surprisingly good recovery from surgery. Gastric biopsy showed an eosinophilic granulomatous vasculitic lesion with no evidence of malignancy. There was an eosinophil infiltrate in the mucosa and submucosa with some plasma cells, lymphocytes and many giant cells, some of which had formed into granulomas centred around small and medium-sized blood vessels with destruction of the internal elastic lamina (Figure 1). No polarizing material, or parasites, or malignant cells were seen in multiple sections. No *Helicobacter pylori* was seen. Stool examination was negative for intestinal

parasites and cysts and helminth ova. Indirect fluorescent antibody test for filaria was negative. The perinuclear ANCA serology was positive, but ANA, anti-double-stranded DNA antibodies and HBsAg were negative. Her eosinophil count was repeated six times but was never more than 15×10^9/l.

Figure 1. . High-power view of biopsy and a granuloma showing many eosinophils in the infiltrate (H & E).

A diagnosis of an eosinophilic granulomatous vasculitis, probably a variant of Churg–Strauss syndrome, was made and she was treated with prednisolone 30 mg daily with continued nutritional support. She showed clinical improvement within 2 weeks of starting steroid treatment, and she could stand and walk alone and felt well enough to go home after four weeks. At 6 weeks her chest X-ray showed almost clear lung fields and osteoporosis which was assumed to be due to steroid therapy, and she gained 5.5 kg in weight. However, 10 weeks after she left hospital, she was admitted with bilateral pneumonia with two left lung abscesses due to a multidrug resistant *Klebsiella* species sensitive to netilmicin, which was unresponsive to treatment. A post-mortem showed that the gastric lesion had resolved. Lung histology showed acute inflammation, necrosis and abscess formation compatible with a severe bacterial infection.

Our patient had a histologically proven EGV but did not have a history of allergy, asthma or peripheral blood eosinophilia. A peripheral eosinophilia of more than 1.5×10^9/l bronchial asthma and a history of allergy are characteristic features of CSS.[1, 2] Therefore her condition could be regarded as being an atypical case of CSS or an eosinophilic variant of WG or PAN.[1, 4] In CSS, patients may fulfil the criteria for hypereosinophilic syndrome at various stages in the course of the disease.[1] One patient with CSS is reported to have had gastrointestinal symptoms three years before developing asthma.[3] Non-asthmatic cases of CSS have also been reported recently,[5] thus the absence of asthma should not automatically exclude a diagnosis of CSS.[5] Furthermore, although peripheral blood eosinophilia is a characteristic feature of CSS, only 80% of these patients have eosinophilia at presentation.[1] Our patient did not have any evidence of sinusitis, runts, destructive lesions in the upper respiratory tract or renal involvement. She also had a positive perinuclear ANCA test and the HBsAg was negative. This makes WG or PAN unlikely; these features are compatible with a diagnosis of an atypical CSS.[1, 2, 6]

The unusual feature about our patient was her presentation. The clinical features strongly suggested a gastric carcinoma. In fact the endoscopic appearance, gastrograffin study (Figure 2) and findings at laparotomy also indicated an infiltrative gastric neoplasm. But the endoscopic biopsies were inconclusive. The diagnosis of EGV was made only after a full thickness gastric biopsy was examined, and was subsequently supported by her dramatic response to systemic steroids − with improvement of her symptoms, clearance of the opacities in the chest X-ray and post-mortem findings.

Figure 2. . Gastrograffin meal showing an irregular mass in stomach.

Granulomatous diseases, including EGV, may rarely mimic infiltrative lesions of the stomach. Establishment of the diagnosis may be difficult, as superficial biopsies of the gastric mucosa may be

normal. Aggressive investigation with full thickness surgical biopsies may be needed to establish the diagnosis and may spare the patient unnecessary radical surgical procedures. Steroid therapy and other immunosuppressive drugs are the treatment of choice in these cases.[1, 2, 6] However such treatment may be hazardous in these patients as they may be malnourished and debilitated, and prone to life-threatening infections.

References

1 *Fauci AS. The vasculitis syndromes. In Isselbacher KJ, Braunwald E, Wilson JD, Martin JB, Fauci AS, Kasper DL eds. Harrison's Principles of Internal Medicine, 13th edn. New York: McGraw-Hill, 1994.*

2 *Lhote F, Guillevin L, Godeau P. Allergic angiitis with granulomatosis: the Churg–Strauss syndrome. Rev. Med. Interne 1994;* ***15*** *(Suppl. 2); 226s 233s.*

CrossRef (/whalecomonlinelibrary.wiley.com/whalecom0/resolve/reference/XREF?id=10.1016/S0248-8663(05)82240-4), PubMed (/whalecomonlinelibrary.wiley.com/whalecom0/resolve/reference/PMED?id=80790

(?url_ver=Z39.88-2004&rft_val_fmt=info%3Aofi%2Ffmt%3Akev%3Amtx%3Ajournal&rft.genre=article&rft.jtitle=Rev.%20Med.%20Interne&rft.atitle=Allergic%20angiitis%20with%20granulomatosis%3A%20the%20Churg%E2%80%93Strauss%20syndrome.&rft.volume=15&rft.issue=Suppl.%202&rft.spage=226s&rft.epage=233s&rft.date=1994&rft.aulast=Lhote&rft.aufirst=F&rfr_id=info%3Asid%2Fwiley.com%3AOnlineLibrary)

3 *Fraioli P, Barberis M, Rizzato G. Gastrointestinal presentation in Churg–Strauss syndrome: a case report. Sarcoidosis 1994;* ***11;*** *42 45.*

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(?url_ver=Z39.88-2004&rft_val_fmt=info%3Aofi%2Ffmt%3Akev%3Amtx%3Ajournal&rft.genre=article&rft.jtitle=Sarcoidosis&rft.atitle=Gastrointestinal%20presentation%20in%20Churg%E2%80%93Strauss%20syndrome%3A%20a%20case%20report.&rft.volume=11%3B&rft.spage=42&rft.epage=45&rft.date=1994&rft.aulast=Fraioli&rft.aufirst=P&rfr_id=info%3Asid%2Fwiley.com%3AOnlineLibrary)

4 *Shimamoto Q, Hirata I, Ohshiba S, Fqjiwara S, Nishio M. Churg–Strauss syndrome (allergic granulomatous angiitis) with peculiar multiple colonic ulcers. Am. J. Gastroenterol. 1990;* ***85;*** *316 319.*

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(?url_ver=Z39.88-2004&rft_val_fmt=info%3Aofi%2Ffmt%3Akev%3Amtx%3Ajournal&rft.genre=article&rft.jtitle=Am.%20J.%20Gastroenterol.&rft.atitle=Churg%E2%80

%93Strauss%20syndrome%20%28allergic%20granulomatous%20angiitis%29%20with%20peculiar%20multiple%20colonic%20ulcers.&rft.volume=85%3B&rft.spage=316&rft.epage=319&rft.date=1990&rft.aulast=Shimamoto&rft.aufirst=Q&rfr_id=info%3Asid%2Fwiley.com%3AOnlineLibrary)

5 *Chen KR, Ohata Y, Sakurai M, Nakayama H. Churg–Strauss syndrome: Report of a case without pre-existing asthma. J. Dermatol. 1992;* ***19;*** *40 47.*

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(?url_ver=Z39.88-2004&rft_val_fmt=info%3Aofi%2Ffmt%3Akev%3Amtx%3Ajournal&rft.genre=article&rft.jtitle=J.%20Dermatol.&rft.atitle=Churg%E2%80%93Strauss%20syndrome%3A%20Report%20of%20a%20case%20without%20pre%E2%80%90existing%20asthma.&rft.volume=19%3B&rft.spage=40&rft.epage=47&rft.date=1992&rft.aulast=Chen&rft.aufirst=KR&rfr_id=info%3Asid%2Fwiley.com%3AOnlineLibrary)

6 *Manganelli P, Trosie Riode W, Buzio C, Pavesi G, Gemignani F. Churg–Strauss syndrome: personal case load and review of the literature. Minerva Med. 1994;* ***85;*** *387 393.*

PubMed (/whalecomonlinelibrary.wiley.com/whalecom0/resolve/reference/PMED?id=7936357)

(?url_ver=Z39.88-2004&rft_val_fmt=info%3Aofi%2Ffmt%3Akev%3Amtx%3Ajournal&rft.genre=article&rft.jtitle=Minerva%20Med.&rft.atitle=Churg%E2%80%93Strauss%20syndrome%3A%20personal%20case%20load%20and%20review%20of%20the%20literature.&rft.volume=85%3B&rft.spage=387&rft.epage=393&rft.date=1994&rft.aulast=Manganelli&rft.aufirst=P&rfr_id=info%3Asid%2Fwiley.com%3AOnlineLibrary)

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