

## CASE REPORTS

## Phlegmonous gastritis: an unusual presenting symptom of Sjögren's syndrome

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### Abstract

**This case report describes the histological and macroscopic changes seen within a few months in the gastric mucosa of a 28 year old woman patient with upper abdominal symptoms. With hindsight these changes were the first signs of Sjögren's syndrome.**

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The Sjögren syndrome is a chronic inflammatory autoimmune disorder usually affecting middle aged women.<sup>1,2</sup>

The syndrome is either primary or secondary. The secondary type is associated with other autoimmune diseases such as rheumatoid arthritis or systemic lupus erythematoses. The damage to the exocrine glands is as a result of infiltration and destruction of the gland tissue by lymphocytes. This is shown clinically by the decrease or total loss of secretion of the affected glands.

Many organs can be affected in the disease. Lymphocytic infiltration can occur in all exocrine glands, including those of the gastrointestinal mucosa.<sup>3</sup> We describe a patient with Sjögren's syndrome and an uncommon gastric mucosal lesion shown to be a phlegmonous gastritis. This phlegmonous gastritis evolved later into a chronic atrophic gastritis, which has

frequently been reported as a symptom of Sjögren's syndrome.<sup>4</sup>

### Case history

A 28 year old Turkish woman was admitted to our ward complaining of severe upper abdominal pain. She also reported anorexia and nausea without vomiting. One year earlier she had been seen in our hospital for joint pain and skin lesions on the lower legs which resembled erythema nodosum. At that time a diagnosis of sarcoidosis was considered but could not be proved. No specific treatment was given and her symptoms subsided spontaneously. Before this admission she had taken no drugs.

Physical examination showed no abnormalities. Laboratory results showed an increased erythrocyte sedimentation rate of 44 mm in the first hour, a raised C reactive protein value of 49 mg/l. The angiotensin converting enzyme activity in serum was normal. A gastroscopy showed a severely swollen, red mucosa on which a large number of small white fibrin dots were seen, the macroscopic aspect resembling 'raw minced meat' (Fig 1). The lesions extended into the duodenum beyond the papilla of Vater. Extensive and deep biopsy specimens were taken, the gastric mucosa felt infiltrated at biopsy. The endoscopic differential diagnosis comprised lymphoreticular malignancy, such as  $\alpha$  chain disease or a non-Hodgkin's lymphoma, while clinically sarcoidosis was considered because of the previous medical history. In the gastric mucosal biopsy specimens a severe phlegmonous inflammation was found. A cellular infiltrate, containing neutrophilic and lymphoid cells with fields of plasma cells that were located in and around the glandular tissue was found, causing glandular destruction. Crypt abscesses were also seen (Fig 2).

A histological diagnosis of phlegmonous gastritis was made. This is known to be a life threatening condition and antibiotic treatment was started while awaiting bacterial cultures of the biopsy specimens and gastric contents. Firstly amoxicillin, then cefuroxim and metronidazol were given for 14 days. During this antibiotic treatment the patient experienced no relief of symptoms. A repeated endoscopy and biopsy specimens of the gastric lesions showed no change. Multiple cultures (of the gastric contents and gastric mucosa) showed no growth of any micro-organism. During antibiotic treat-

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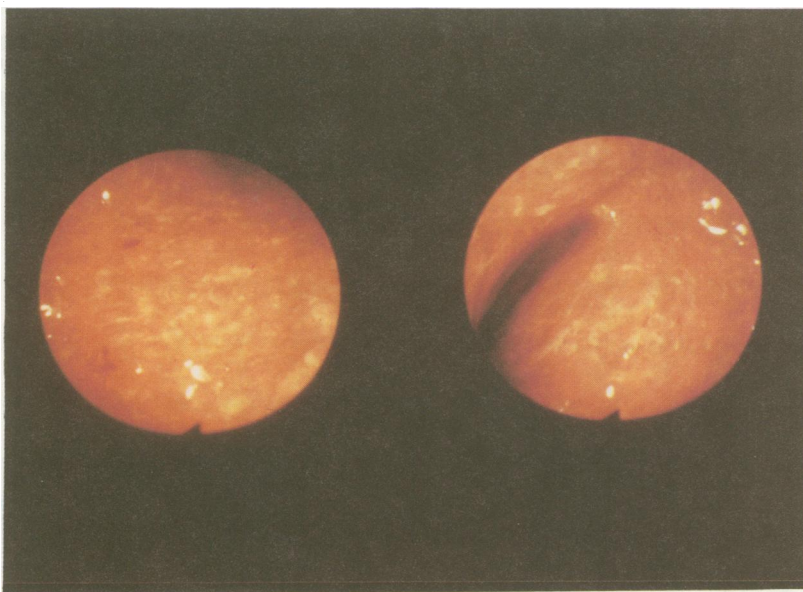


Figure 1: Macroscopic aspect of the gastric mucosa seen during the first gastroscopy.

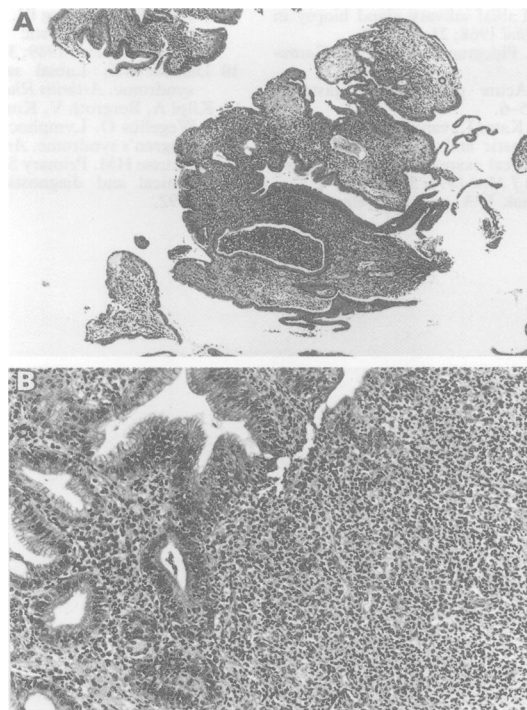


Figure 2: Histological examination of the gastric mucosa taken of the area shown in Fig 1. In (A) a crypt abscess can be seen and in (B) the mixed nature of the inflammatory infiltrate.

ment the patient developed a severe generalised rash as an allergic reaction for which 30 mg prednisolone was given. Both the rash and the gastric symptoms disappeared immediately after corticosteroid treatment.

A somatostatin scan was performed as part of an ongoing research project because the diagnosis sarcoidosis was considered. This scan showed a high uptake of radioactive labelled somatostatin in the areas around the eyes and parotid glands, no uptake being seen in the gastric region. These typical localisations suggested Sjögren's syndrome. Specific questioning showed symptoms of dry eyes and dry mouth. The Schirmer and break up time test confirmed a decrease of tear secretion. A sublabial gland biopsy showed more than three lymphocyte foci per 4 mm<sup>2</sup> confirming Sjögren syndrome according to the criteria of Chisholm and Mason.<sup>5</sup> A pentagastrine test showed the presence of achlorhydria. Serological indices, such as anti-nuclear antibodies, rheumatoid factor, antibodies to SS-A and SS-B, and also antibodies against parietal cells and intrinsic factor were negative. The steroid treatment was continued, with 30 mg prednisone orally once every day.

A gastroscopy two months later showed an extensive atrophic gastritis. The signs of inflammation had vanished. Histological examination confirmed a severe chronic atrophic gastritis.

One and a half years later gastroscopy showed some slight fasting gastric secretion. In the fundus and corpus of the stomach an increase in superficial vascular pattern was visible. The mucosa at the small curvature and at the antrum still showed some areas with slightly red swollen mucosa containing white 'point like' fibrin patches. Histological examination of these areas at this time confirmed a chronic gastritis and

atrophic gastric mucosa. The patient was taking 5 mg prednisone and had no complaints.

### Discussion

The Sjögren or Sicca syndrome usually presents clinically with symptoms of a dry mouth and dry eyes. These phenomena are a result of infiltration by lymphocytes of the affected exocrine glands.

The initial symptom in our patient was upper abdominal pain. A gastroscopy showed a severe inflammation of the stomach and proximal duodenum. Histological examination showed what seemed to be a life threatening phlegmonous gastritis. In the same biopsy infiltration of the mucosal stroma by a large number of lymphocytes and plasma cells were seen. The differential diagnosis of phlegmonous gastritis consists of bacterial infections especially *Staphylococcus aureus* and including tuberculosis.

Bacterial phlegmonous gastritis either responds to antibiotic treatment or results in death of the patient.<sup>6,7</sup> In our patient all cultures were negative and antibiotic treatment had no effect. Other diseases such as sarcoidosis, lymphoreticular malignancies, Crohn's disease, and non-steroid anti-inflammatory drug induced gastropathy were ruled out in our patient.

The diagnosis of primary Sjögren's syndrome was finally made on the typical symptoms of dry eyes, dry mouth, the abnormal Schirmer test, and a positive labial salivary gland biopsy.<sup>8-10</sup> In this patient Sjögren's syndrome presented with a phlegmonous gastritis that responded promptly to steroid treatment. To our knowledge this presentation has not been described previously. Gastrointestinal involvement is reported to occur frequently in the Sjögren syndrome. Some authors have described an incidence of chronic atrophic gastritis in more than 65%<sup>11</sup> of all cases of Sjögren's syndrome. The macroscopic aspect of the stomach is described as atrophic or having a cobble stone aspect.<sup>4,8</sup> Immunohistological investigation shows the same changes in the gastrointestinal mucosa as seen in the salivary glands.<sup>5</sup>

The positive somatostatin scan was caused by a high expression of somatostatin receptors on the lymphocytes infiltrating the salivary and tear glands. This phenomenon has been found in some patients with Sjögren's syndrome in our hospital (unpublished data). This deserves further investigation, especially because salivary scintigraphy using technetium-99m pertechnetate in patients with proved primary Sjögren's syndrome is of limited discriminative value.<sup>12</sup> In conclusion phlegmonous gastritis was found as the presenting symptom in a case of Sjögren's syndrome and reacted well to steroid treatment. An atrophic gastritis, which is a more usual finding in Sjögren's syndrome, developed subsequently.

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