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Gastric sarcoidosis — a case report

Abstract

Gastrointestinal sarcoidosis is very rare manifestation of the illness. This article presents patient who had been diagnosed for several months because of abdominal pain, vomiting and significant weight loss. Numerous gastric endoscopy examinations showed the difficult healing ulcers in the stomach, progressive thickening of the mucosal folds with narrowing of the lumen, lack of peristalsis and the rigidity of the walls. Histological examination of the specimen of the ventricular mucosa revealed chronic inflammation, as well as suspicion of tumor infiltration. Finally gastric sarcoidosis has been diagnosed on the basis of CT with double contrast, and histopathological examination of biopsis of stomach collected during laparotomy.

Key words: sarcoidosis, gastric sarcoidosis

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Introduction

Sarcoidosis is a disease of unknown aetiology characterised by the formation of noncaseating granulomas localised in the lungs in more than 90% of cases [1]. Extrapulmonary location of sarcoid granulomas is also possible, but gastrointestinal sarcoidosis is very rare. It is estimated that this location with the associated symptomatology is observed in about 0.6–1% of sarcoidosis patients [2, 3]. However, it has been suggested that sarcoid granulomas may be observed in the stomach of as many as 10% of patients with pulmonary manifestation of this disease without causing any significant gastrointestinal complaints [4].

We present the difficulties encountered in the attempts to establish the cause of abdominal pain in a patient subsequently diagnosed with gastric sarcoidosis. This, according to our best knowledge, is the first case of gastric sarcoidosis reported in Poland.

Case presentation

A 43 year-old male welder (a 20 cigarettes a day smoker) was admitted to the Department of Lung Diseases and Tuberculosis in September 2008 complaining of epigastric pain, periodic vomiting and weight loss. In 2005, based on the histopathological examination of tissue samples collected during right-sided thoracotomy, the patient had been diagnosed with pulmonary sarcoidosis. Radiological studies performed at that time revealed hilar lymphadenopathy and disseminated finely mottled opacities in both lungs (Fig. 1) and the patient was then managed with prednisone at an initial dose of 30 mg for one year, after which regression of the pulmonary lesions was observed.

In June 2008, the patient developed epigastric pain which intensified after meals and was accompanied by vomiting and weight loss. A gastrointestinal panendoscopy carried out the following month revealed erosions in the oesophagus and an irregular ulceration of 30 mm in diameter in the gastric body in the anterior wall and erosive inflammation of the mucous membrane lining the body, cardia and duodenal bulb. Histopathological examination of the tissue samples collected from the ulceration area revealed a chronic ulcer. Despite treatment with proton pump inhibitors, the symptoms failed to subside and further weight loss was

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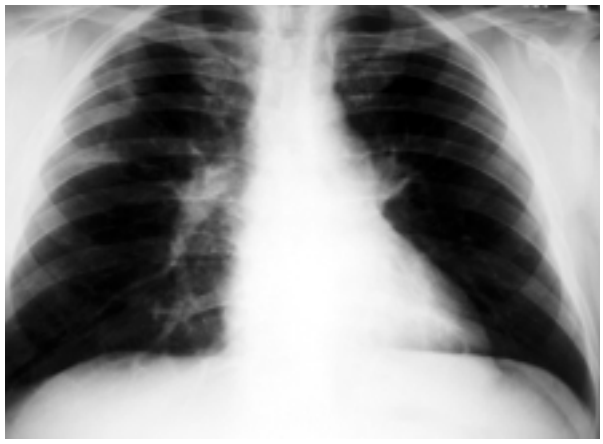

Figure 1. Chest X-ray taken in February 2005: Hilar lymphadenopathy and disseminated lesions in both lungs

observed. A follow-up endoscopy two weeks later revealed progression. In addition to the persistent ulceration in the gastric body, an extensive ulceration in the antrum was demonstrated, which included all its periphery and the pylorus. Other findings included a narrowing of the lumen of the prepyloric region, poor distensibility of the gastric wall following air insufflation and no peristalsis. Histopathological examination of the tissue samples collected from the ulcerations revealed chronic gastritis and intestinal metaplasia. The antiulcer treatment was intensified (omeprazole 20 mg BID and famotidine 40 mg at bedtime). The next endoscopy carried out in September 2008 revealed a nearly complete healing of the ulcers but at the same time demonstrated poorly visible peristalsis, narrowing of the stomach, thickening of the folds and lack of distensibility with insufflation of air. Again, tissue samples for histopathology were collected (as a malignancy was suspected).

The histopathological examination revealed a microcellular infiltration of a probably malignant nature. The patient was consulted at the Oncology Institute in Gliwice. Immunohistochemistry did not confirm the malignant nature of the lesions. Because the symptoms persisted and the patient continued to lose weight (about 30 kg over 4 months) another panendoscopy was performed, with a similar result to the first one. This time, however, based on the histopathological examination (granulomatous chronic gastritis), a working diagnosis of suspected sarcoidosis was made and the patient was referred to the Department of Lung Diseases in Zabrze. An X-ray film, obtained in June 2008 and provided with the patient, did not show any changes.

On admission, the patient complained of epigastric pain, a feeling of abdominal fullness after meals and periodic throwing up of food. The haematological and biochemical parameters of his blood were all normal.

During hospitalisation, the patient underwent an abdominal CT scan following oral administration of 3% aqueous solution of a contrast medium, and before and after intravenous administration of the contrast medium. The scan revealed thickening of the gastric wall consistent with intramural infiltration (linitis plastica-type lesions) and intra- and retroperitoneal lymphadenopathy (Fig. 2).

The patient was transferred to the Clinical Ward of General and Bariatric Surgery and Emergency Medicine at the Specialist Hospital in Zabrze with suspected gastric sarcoidosis requesting verification of the diagnosis. On 26 January 2009, the patient underwent a laparotomy with excision of the lymph node in the region of the hepatoduodenal ligament and collection of a full-thickness gastric wall tissue sample from the body of the stomach. The histopathological examination of the lymph node revealed "a picture of granulomatous inflammation that could be consistent with sarcoidosis". The patient was re-hospitalised at the Department of Lung Diseases, where the eradication of *Helicobacter pylori* (omeprazole 20 mg BID, metronidazole 500 mg BID, amoxicillin 1,000 mg BID for 14 days and prednisone at the initial dose of 60 mg) was started. As a result of the treatment, the patient's condition improved, the epigastric pain subsided and he gained 10 kg in weight. A follow-up

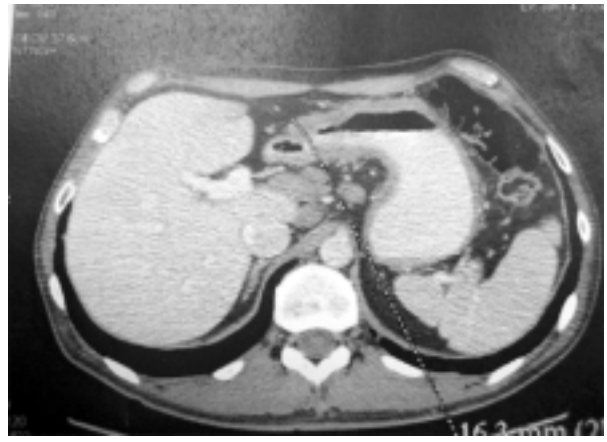

Figure 2. Abdomen CT from 14 January 2009:

Stomach walls evidently thickened: from the fundus in direction of the pylorus the wall thickness progressively increases, reaching maximally 16.3 mm. In multiphase scan x-ray attenuation by thickened walls tends to be lower than normal. Multiple enlarged lymphnodes located intra and extraperitoneally (maximally 22 × 19 mm), lymphnodes located above left renal vein opening to superior vena cava. Conclusion: Enlarged intra and extraperitoneal lymphnodes. Thickened stomach walls suggesting intramural infiltration

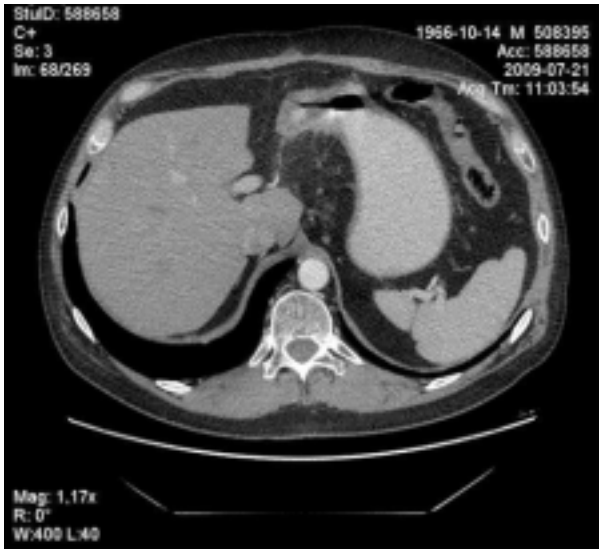

Figure 3. Abdomen CT from 21 July 2009:

Slightly thickened stomach wall — maximally anterior wall measures 6 mm and in right-side recumbent position, wall thickness in the pre-pyloric region reaches 9 mm. X-ray attenuation of the stomach wall is normal. Intraperitoneal lymphnodes present: along minor curvature of the stomach — maximal dimension 10.7 × 16.3 mm, peripyloric lymphnode 9.8 × 7.4 mm, pancreatoduodenal 8.6 × 9.5 mm, above renal vein lymphnode measuring 13.5 mm.

Conclusion: Compared with the previous CT dated 14 Jan 2009 a significant regression is noted in both: stomach wall thickness and lymphnodes dimensions. Currently the largest lymphnode is located in stomach minor curvature

abdominal CT scan after six months of prednisone treatment revealed a considerable regression of lesions both in the gastric wall and in the lymph nodes (Fig. 3).

The patient remains under constant monitoring and is still being treated with prednisone at the dose of 20 mg and omeprazole at the dose of 20 mg QD. A further weight gain of 10 kg has been observed. The patient reports no gastrointestinal complaints.

Discussion

Sarcoid granulomas have been observed in most organs of the gastrointestinal tract: namely in the salivary glands, oral cavity, lips, oesophagus, stomach, small intestine, colon, appendix, peritoneum, liver, spleen, pancreas, bile ducts and gallbladder [2, 5]. However, apart from the liver, the stomach is the organ most commonly affected by the sarcoid process [2, 3, 5].

A systematic review of gastrointestinal sarcoidosis cases indicates that more than 60 cases of gastric sarcoidosis had been reported since the early 1990s [2].

The clinical manifestation of gastric sarcoidosis varies greatly and, obviously, is very non-specific [2, 5–7]. Patients may complain of epigastric pain, nausea, vomiting or weight loss. Weight loss may be considerable and suggest gastric cancer. Epigastric pain most commonly appears after meals and is described as burning, dull or cramping. The clinical signs and symptoms of gastric sarcoidosis may suggest cancer, hypertrophic gastritis or Crohn's disease, but above all peptic ulcer with gastrointestinal bleeding, secondary anaemia or pylorostenosis. Rarely, massive gastrointestinal bleeding occurs [8, 9]. Such sudden and massive bleeding is the sole manifestation of gastric sarcoidosis and leads to death [10]. Co-existent hypersplenism and thrombocytopenia may predispose to bleedings from a sarcoid gastric ulcer [3].

It is estimated that the predominant symptom is bleeding in 25% and pain in 75% of patients [11]. Oesophagitis and gastrooesophageal reflux may occur in 21% of patients [1]. There has been a report of a case of gastric sarcoidosis whose clinical manifestation resembled that of irritable bowel syndrome [12].

The sarcoid lesions in the stomach may also be accompanied by the presence of sarcoid granulomas in the liver and spleen and in retroperitoneal lymph nodes [6]. Hilar lymphadenopathy and/or disseminated lesions in the lungs may co-exist with, or precede, the development of sarcoid lesions in the gastrointestinal tract [6, 7, 13]. This was the case in our patient. Isolated gastric sarcoidosis is very rare [2, 3].

Gastric sarcoidosis may not cause any significant symptoms and then the disease is suspected based on the results of histopathological examination of biopsies collected during flexible gastroscopy [4, 14]. Chlumsky et al. [14] found sarcoid granulomas in biopsies collected during routine upper gastrointestinal endoscopies in 0.02% of 500 patients.

Gastroscopy may reveal numerous ulcers, mainly in the cardia [9]. Sometimes areas of mucosal reddening, swelling or atrophic and/or polypous lesions are visible in the cardia [12, 13]. Numerous nodular lesions in the mucous membrane in the region of the lesser curvature near the cardia are also sometimes observed [13]. The gastric lumen is often narrowed, which is relatively easy to see in single-contrast X-ray studies, which can also reveal ulcer niches [15]. In gastric sarcoidosis, X-ray can also demonstrate a conical, smooth narrowing of the cardia. Less frequently, it may reveal irregular narrowing of the entire stomach resembling linitis plastica seen in gastric cancer

[15]. Double-contrast X-ray studies sometimes reveal irregular folds and nodular lesions of the mucous membrane [15].

Taking into account the above pathological changes in gastric sarcoidosis, four principal categories of lesions have been distinguished: subclinical (the commonest), ulcerative, infiltrative and polypous [3].

The diagnosis of gastric sarcoidosis may be made on the basis of histopathological evaluation of biopsies collected during endoscopy, but similar microscopic pictures are observed in Crohn's and Whipple's diseases. Differential diagnosis must also include tuberculosis, fungal infections and sarcoid reactions in cancer, which we took into account in the evaluation of our patient. Forty per cent of patients may have a co-existent *Helicobacter pylori* infection [2]. It should be remembered that endoscopic biopsies sometimes fail as regards the identification of sarcoid lesions, as the granulomas may be localised in the submucosa and the deeper layers of the gastric wall, not just in the mucous membrane.

The prognosis in gastrointestinal sarcoidosis, including gastric sarcoidosis, is generally good, with glucocorticosteroids being the treatment of choice [5, 14]. Two thirds of patients considerably improve on glucocorticosteroids, although the clinical improvement is not always paralleled by a resolution of pathological lesions [11]. Surgery is necessary in rare cases with massive bleeding, pylorostenosis or suspected malignancy based on radiological or endoscopic studies [2, 3, 5].

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