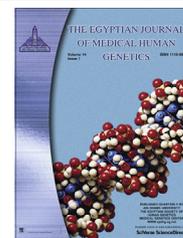




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CASE REPORT

Gastric sarcoidosis in children: A case report and review of the literature

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KEYWORDS

Gastric sarcoidosis;
Corticosteroid therapy;
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Abstract Sarcoidosis is a systemic granulomatous disease of unknown etiology that is characterized by the formation of noncaseating granulomas. Gastrointestinal (GI) tract involvement in sarcoidosis is rare. Gastric sarcoidosis, particularly involving the antrum, affects approximately 10% of patients with systemic disease. GI sarcoidosis commonly occurs subclinically, with clinical manifestations present in only 0.1–0.9% of patients with the disease.

This is a rare case report of a 8 year Saudi girl with symptomatic gastric sarcoidosis. The patient presented with anorexia, postprandial upper abdominal pain and fullness, and weight loss of 3 months duration. She was presented acutely after 6 months with attack of hematemesis. Endoscopic examination of upper gastrointestinal tract revealed bleeding nodular mucosal irregularities. Mucosal biopsies revealed noncaseating granulomatous inflammation involving the gastric mucosa confirming the diagnosis of sarcoidosis. Corticosteroid therapy was started and the symptoms abated almost immediately. We also offer a review of the literature.

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Introduction

Sarcoidosis is a disease of undetermined cause that primarily affects adults and is characterized by the formation of noncaseating granulomas. Proper interpretation of the biopsies is crucial because tuberculosis, syphilis, Crohn disease, foreign body reactions, and fungal infections can all present with

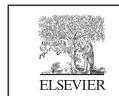
granulomatous disease [1]. Although sarcoidosis is a multisystem disease, involvement of alimentary tract is rare; when present, it is often an asymptomatic finding in a patient with widely disseminated disease. The etiology of sarcoidosis is unknown, most experts think that sarcoidosis results from the exposure of genetically susceptible hosts to specific environmental, occupational or infectious agents that trigger an exaggerated cellular immune response, leading to granuloma formation. Follow-up studies on chromosome 6 identified the *BTNL2* gene to be associated with sarcoidosis [2].

This is a rare case report of a 8 year Saudi girl with symptomatic gastric sarcoidosis presented with anorexia, upper abdominal pain, weight loss of 3 months duration, and 6 months later presented with hematemesis and abated almost immediately with corticosteroid treatment.

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Case presentation

A Saudi girl (8 years old) presented with 3 month history of anorexia, post prandial upper abdominal pain and fullness. She was also complaining of generalized fatigue, early satiety, paleness and weight loss of 5 kg in last 3 months. She denied experiencing diarrhea, fever, rashes, joint pain, or respiratory complaint. Her past medical history was unremarkable except attacks of mild epistaxis. Her parents are nonconsanguineous with no family history of gastrointestinal, food allergy autoimmune, tuberculosis, or malignant diseases.

On physical examination, she looked pale, no jaundice, her weight on 25% centile, height on 50% centile, hepatomegaly 4 cm below costal margin. Vital signs were normal, no skin rashes, no palpable lymph nodes and normal chest and neurological examinations. The investigations showed: WBCs $12.4 \times 10^9/l$, neutrophils 35%, the Hb level was 6.4 gm/dl, MCV 51%, RDW 17, retics 1.5%, platelets $520 \times 10^9/l$, Coombs test -ve, Hb electrophoresis was normal, Iron profile: iron 1.5 $\mu\text{mol/l}$, transferrin 2.7 $\mu\text{mol/l}$ (N: 2.1–3.6), saturation 2%, CRP -ve, ESR 50 mm/h, stool occult blood -ve. Serum electrolytes liver, renal, lipid profiles were normal, CXR showed bilateral basal infiltrates with minimal left pleural effusion. Ultrasonography of abdomen showed hepatomegaly with iso-hypoechoic lesions in liver suggesting granulomatous disease as the primary possibility over malignant process, C.T chest and abdomen showed multiple small mediastinal lymph nodes; multiple small subpleural nodules scattered at both lungs, hepatosplenomegaly with multiple small innumerable lesions at the liver. Tuberculin skin test was 9 mm, sputum and gastric aspirate for acid fast bacilli stain and cultures were -ve, sweat test was 10 mm, immunoglobulin electrophoresis; normal, antibacterial antibodies within normal, nitroblue test; normal, C50/100 normal, ANA -ve, Hbs Ag; -ve. Bone marrow examination showed normal cellularity with active trilineage hematopoiesis. Liver biopsy showed nonspecific granulomatous inflammatory reactions. She was put on antituberculous therapy for 3 months without improvement in symptoms.

She was presented acutely after 6 months with an attack of hematemesis needing blood transfusion. Urgent endoscopic examination of upper GIT showed nodular mucosal irregularities in the fundus of stomach; esophagus, antrum, pylorus and duodenum were normal. Mucosal biopsies revealed multiple non caseating granulomas confirming the diagnosis of sarcoidosis. Stains for *Helicobacter pylori*, acid fast, and fungi were negative, serum angiotensin converting enzyme was 74 units/l (N 44–125 units/L), 24 h urine collection ruled out hypercalciuria. She was treated with prednisolone, metoclopramide, and antacid and obtained relief of gastric symptoms, noted weight gain and improvement in her energy level. The dose of prednisolone was gradually tapered to a maintenance dose of 10–15 mg over a period of approximately 6 months, upper gastrointestinal barium study performed later showed a decrease in gastric nodularity.

Discussion

Sarcoidosis is a multisystem systemic granulomatous disease of unknown etiology that most commonly affects young adults. The disease is relatively rare in the pediatric population.

The true incidence and prevalence of childhood sarcoidosis is unknown because of the rarity of the disease and the small number of reported cases in childhood.

Two distinct forms of childhood sarcoidosis appear to exist. Older children usually present with a multisystem disease similar to the adult manifestation with frequent lymphadenopathy and pulmonary involvement, as well as generalized signs and symptoms, such as fever, malaise, and weight loss. In contrast, early-onset childhood sarcoidosis is a unique form of disease characterized by the triad of rash, uveitis, and arthritis in children who are younger than 5 years [3].

The lung is the organ most commonly involved in sarcoidosis. Pulmonary symptoms are usually mild and often consist of a dry hacking cough, with or without dyspnea. Bilateral hilar lymphadenopathy with or parenchymal involvement is the most common radiographic finding as in our case [4].

Peripheral lymph node enlargement noted in 40–70% of cases, is the most accessible site for diagnostic biopsy. Lymph nodes are typically firm, non-tender, and freely movable. Although hepatosplenomegaly may occur in up to 43% of patients with childhood sarcoidosis, clinical manifestations are not as apparent [5].

Ocular involvement is common in childhood sarcoidosis. Visual symptoms such as eye pain, blurry vision, photophobia, and redness may be present in 29% of patients. Uveitis or iritis is the most common manifestation of the children with sarcoidosis. Slit lamp examination is mandatory in the evaluation of childhood sarcoidosis [6].

An erythematous rash is commonly noted in childhood sarcoidosis. The most frequent cutaneous eruptions include soft, red to yellowish brown, or violaceous, flat-topped papules, found most frequently on the face. Macular lesions with scarring and ichthyosiform cutaneous manifestations are frequently encountered [7].

Gastrointestinal tract sarcoidosis is rare and may present in the context of generalized disease as in our case or as an isolated finding. GI sarcoidosis commonly occurs subclinically, with clinical manifestations present in only 0.1–0.9% of patients with the disease. Manifestations of the disease include epigastric pain, nausea, vomiting, and are usually postprandial. Weight loss is common, upper GI bleeding can also be the initial presentation. Endoscopy may reveal ulcerations, gastritis, or diffusely erythematous, friable mucosa that is elevated over the surroundings. Nodular irregularities and mucosal polyps may be present occasionally. The diagnosis of gastric sarcoidosis is dependent on histological evidence of sarcoid granulomas on mucosal biopsies [8].

There is no laboratory test diagnostic of sarcoidosis. Laboratory evaluation may reveal elevated erythrocyte sedimentation rate (ESR) or other acute phase reactants. Anemia, leucopenia, and eosinophilia are commonly seen on blood counts. Hypercalcemia and/or hypercalciuria may be found. The serum level of angiotensin-converting enzyme (ACE) is elevated in over 50% of children with late-onset sarcoidosis [9]. Chest radiograph is very useful and may reveal bilateral hilar adenopathy. High-resolution chest computed tomography (CT) can be helpful in delineating the extent of parenchymal disease and hilar adenopathy. The diagnosis of sarcoidosis is confirmed by demonstrating a typical noncaseating epithelioid cell granuloma on a biopsy specimen.

The current therapy of choice for childhood sarcoidosis with multisystem involvement is corticosteroids. Oral prednisolone

is usually initiated at 1–2 mg/kg/d for 4–8 weeks as induction treatment. This treatment is continued until the clinical manifestations of the disease resolve or show significant improvement, then slowly tapered over a period of 2–3 months to an appropriate maintenance dose (10–15 mg per day as a once daily regimen) that is usually required for at least 6 months for most age groups [10].

The disease activity is monitored clinically, radiographically, and by serum ACE levels. Some patients may relapse, either during steroid taper or after discontinuation of the drug. In such cases, steroid treatment should be restarted with a dosage similar to that used in the induction treatment. The overall prognosis of childhood sarcoidosis is reported as good compared with the prognosis for adults. Sarcoidosis in very young children with involvement of eyes, joints and skin has a guarded prognosis with the likelihood of a chronic progressive course [11].

Conclusion

Gastric sarcoidosis should be considered in patients with sarcoidosis who have gastric symptoms. Because of the subclinical nature of disease, it may actually be much more common than thought. It is therefore essential that histologic confirmation be established so that symptoms may be treated appropriately as they arise.

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