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Rare Gastric Granulocytic Sarcoma: A Case Report

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Abstract **Case Report**

Introduction: GS is often composed of myeloblasts with or without signs of maturation of promyelocytes and mesangocytes. The organizational structure is partially or completely destroyed. GS can occur in various parts of the body but gastrointestinal involvement is rare. ***Case presentation:*** An 11-year-old male patient presented with symptoms of systemic blood stasis, fatigue, and abdominal pain as the first symptom. He was diagnosed with acute myeloid leukemia. After treatment, the symptoms were relieved and abdominal pain disappeared. After 9 months, the patient developed abdominal pain again, and was diagnosed as gastric granulocytic sarcoma by ultrasound gastroscopy, pathological examination and immunohistochemical staining. ***Conclusion:*** For patients with AML, whether in the early diagnosis or during the course of chemotherapy, abdominal symptoms should be routinely checked for B-ultrasound and CT. If necessary, gastroscopy and pathological biopsy should be performed to avoid GS misdiagnosis.

Keywords: Granulocytic sarcomas (GS), leukemia, Immunohistochemical staining, case report.

INTRODUCTION

Granulocytic sarcomas (GS) is a tumorous mass formed by extramedullary infiltration and hyperplasia of immature granulocytes [1]. GS is often composed of myeloblasts with or without signs of maturation of promyelocytes and mesangocytes. The organizational structure is partially or completely destroyed. GS can occur in various parts of the body, usually involving lymph nodes, bones, skin, brain, meninges, eyelids, endocardium, peritoneum, breast, liver, kidney, genitals (testicles, ovaries), muscles, etc. But gastrointestinal involvement is rare. GS is primarily associated with acute myeloid leukemia (AML). It usually occurs either during the initial course of AML or after remission has been achieve [2, 3]. This case that was eventually diagnosed as stomach GS by biopsy. But it is not certain whether stomach GS occurred simultaneously with AML or as extramedullary relapse of AML. So, for AML patients, when abdominal pain occurs in the very beginning or during the process of treatment, B ultrasonic, CT, even gastroscopy and pathological biopsy should be received to avoid misdiagnosis GS.

CASE REPORT

In September 2014, an 11-year-old boy, presented with ecchymosis, fatigue and epigastric pain, was admitted to our hospital. There was no history of tuberculosis, hepatitis and previous exposure, neither was family history of blood tumor diseases. Results of blood tests for the first time: hemoglobin 92 g/L, platelet 20x 10^{-9}/ L, neutrophils, lymphocytes, monocytes not detected, white blood cell 20.76 x 10^{-9}/ L, immature cells, primitive granulocyte 70 %. The boy was diagnosed with acute myeloid leukemia (AML-M2) with t (16; 21) (p11; q22) TLS/ERG positive（ intermediate risk. His bone marrow was in remission and fusion gene TLS - ERG turned negative after the patient underwent DA, IA chemotherapy. Further consolidation chemotherapy was given, during which the whole body's ecchymosis, fatigue had been relieved and abdominal pain disappeared.

In June 2015, results of two tuberculosis consecutive bone marrow biopsy showed that the fusion gene TLS-ERG of the boy turned positive, recurrence of molecular was confirmed. Again, dull epigastric pain was increasing gradually without tenderness, rebound tenderness, muscle tension, radiation and so forth. The possibility of acute abdominal disease, such as intestinal

obstruction, perforation and pancreatitis, etc., was excluded by X-ray, CT examination. CT result showed that several local stomach walls, at the bottom of the stomach and the stomach body, appeared to be thickening with coarse edge (Figure-1). In order to make the cause of abdominal pain clear, upper gastrointestinal endoscopic examination were carried out. Whose result showed that at the bottom of the stomach and on the stomach body, there were locally irregular thickening and nodular changing, most of which were along the greater curvature stomach plica (Figure-2). However, considering the specialty of uplift change, ultrasonic gastroscopy was performed to further evaluate the lesion. The result suggested: the local lining of the stomach is clearly visible, submucous and mucous layer thickened obviously while the echo is in a medium and uneven state without vascular, the muscularis propria was still regular (Figure-3). Biopsy was completed under endoscope. Pathologic results showed that the cellular structure was partially damaged, and the infiltration of unusual large cell was visible .One kind of young cells were distributed dispersedly and focally with big cell bodies，abundant cytoplasm，irregular nucleus and visible nucleoli (Figure-4). Immunohistochemical staining results showed that MPO (NS), CD34 (+), CD117 (+), 70 ~ 80 % Ki67 positive rate, LCA (-), the TDT (-). Pathology consultation results from the Peking union medical college hospital was the same as above. Considering that the boy is a patient of acute myeloid leukemia, eventually diagnosis was confirmed as stomach granulocyte sarcoma (leukemia merger myeloid sarcoma).

In October 2015, the definitive diagnosis for the patient was acute myeloid leukemia with partially differentiated type (M2) and t (16; 21) (p11; q22) TLS/ERG positive (intermediate risk), molecular relapse and stomach appeared extramedullary infiltration. The efficacy of the second chemotherapy was not obvious. The patient's family members refused, The patient gave up further treatment because of the refusal of bone marrow transplantation by his relative. We contacted the patient by phone in January 2016 and learned that he had not received any treatment after discharge. At present, the appetite of patients is still poor and relatively stable.

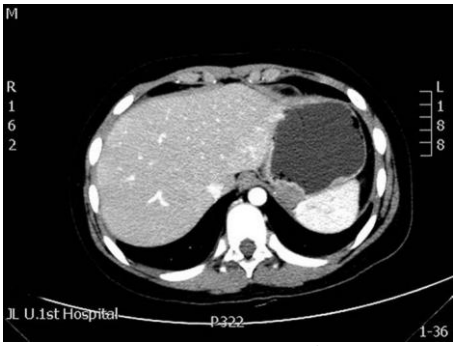

Fig-1: Computerized tomography noted the presence of segmental thickening of the gastric

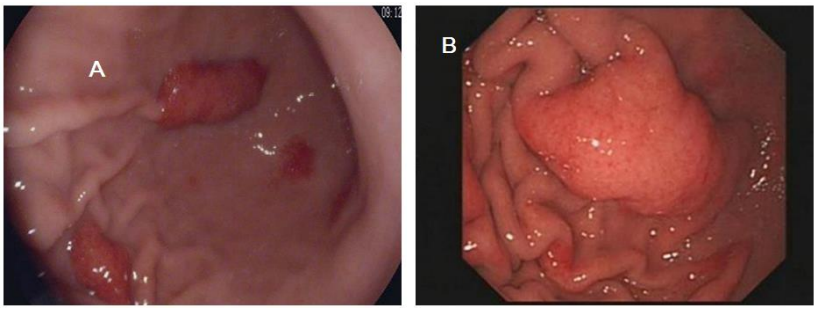

Fig-2: Lesions under the endoscope. (A) Gastric body and gastric antrum (B) Gastric fundus

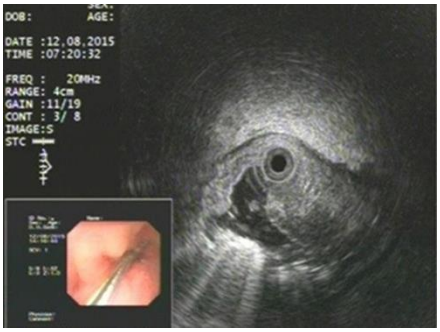

Fig-3: Ultrasonography endoscope result

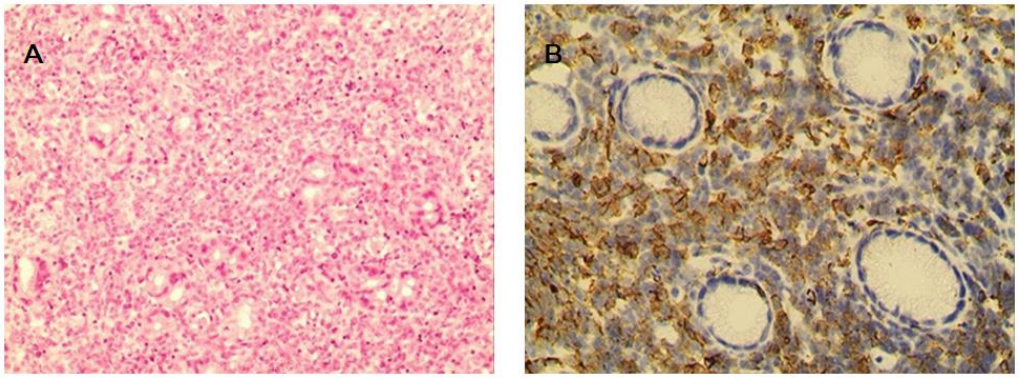

Fig-4: Pathologic examination results: cellular structure partially damaged unusually large cell infiltration is visible (A) H&E staining at a magnification of x10; (B) CD34-immunohistochemical staining was positive

DISCUSSION

Granulocytic sarcomas (GS) are extramedullary tumors of myeloblasts, myeloid precursors and neutrophils in various combinations, and they involve extramedullary blast proliferation from one or more myeloid lineages that replace the original tissue architecture. These tumors are also known as myeloid sarcomas, chloromas or extramedullary myeloid tumors [1]. It is classified as non-leukemia GS (isolation or primary GS) and leukemia GS (leukaemia marrow infiltration GS).The patient in this case belongs to the latter. For acute myeloid leukemia (AML), GS is most frequently associated with French-American-British subtypes M4 and M5, and mainly involves the nervous system [3, 4]. Other article said that in adults GS is most frequently associated with French-American-British subtypes M2 and M4, which is probably one of the most important risk factors for extramedullary involvement in patients with AML [5] The case of this patient is confirmed as AML with maturation (M2), in which extramedullary infiltration rarely occurred to children and AML with gastric involvement is even more rare to see.

GS occurs over a wide range of ages, and it is more common in children than in adults (in some studies pediatric patients have an incidence of up to 30% vs 2%-5% for adult patients) [6]. This case happened to an 11-year-old boy. GS can develop in different sites, such as lymphoid organs, bones (orbit, mastoids, etc.), skin, soft tissues, central nervous system, bladder, breast, and uterus [2]. Gastrointestinal involvement is rare, if GS occurs in this region. The small intestine is a relatively common location for these tumors. The usual presentation includes abdominal pain from a small bowel obstruction. Gastric GS is rarely reported. Only one case of stomach GS was searched in CNKI net and Wanfang database, while no more than 20 cases were found in PubMed database. There aren't any clear guidelines of diagnosis and treatment for GS. Abdominal pain as the initial symptom and severe abdominal pain in the course of the disease for GS patients is rare, and few related cases have been reported in the literature.

GS is closely associated with AML, it usually occurs either during the initial course of AML or after remission has been achieved [7]. The patient with AML was confirmed as stomach myeloid infiltrate after 1-year-chemotherapy. Abdominal pain occurred at the beginning of the disease and relieved after systematic chemotherapy. 9 months later the abdominal pain relapsed, and gradually exacerbated. It was eventually diagnosed as stomach GS by biopsy. There is no endoscopy right at the outset of the disease. Therefore, it is not certain whether stomach GS occurred simultaneously with AML or as extramedullary relapse of AML. So, for AML patients, when abdominal pain occurs in the very beginning or during the process of treatment, B ultrasonic, CT, even gastroscopy and pathological biopsy should be received to avoid misdiagnosis GS.

There is no specific performance under endoscopy for GS. In combination with related literature reports, GS is usually presented in a special local irregular thickening of gastric body and nodular changes, some of them with ulcer on the surface. The ultrasonic shows mucosa and submucosa have changed. Thickening of the submucosa is in a medium uneven echo without vascular. Attention should be paid to identify it among primary tumors of gastrointestinal tract and lymphoma, etc.

Endoscopic biopsy generally gets the superficial layer of the lesion. So the biopsy tissue usually is necrotic exudation and without substantial pathological lesion. It is necessary to get materials from different parts and in a certain depth. For the deep lesions, deep hole type biopsy can be used at the bump. EMR or ESD can be considered to get a bigger tissue when necessary. GS should be given enough attention and biopsy pathology is an important means to diagnose. Early diagnosing should be achieved as much as possible, thus treatment can be performed to prolong disease-free survival.

The histopathologic of GS can be classified into three types according to the degree of myeloid cells differentiation. The first is blastic type, which consists

of archaeocyte; the second is immature type, which consists of archaeocyte and promyelocytic; the third is differentiated type, which consists of promyelocytic and metagranulocyte. In conventional paraffin section, if the tissue form lacks myeloid differentiation and immature eosinophils, besides, cells are immature, diagnosis based on organizational form is easily misdiagnosed as diffuse large B-cell lymphoma, Burkitt lymphoma and lymphoblastic lymphoma, and malignant tumors of non-hematopoietic system, etc [8]. Immunohistochemistry and flow cytometry instrumental technology can dramatically reduce the misdiagnosis rate of GS. In GS immunohistochemical markers, myeloperoxidase (MPO) and the leukocyte differentiation antigen CD34, CD117 sensitivity is high [4, 5].

Traditional treatments for GS include surgery, radiation therapy, chemotherapy, etc. Due to the low incidence of the disease, the diagnosis and treatment experience for GS are relatively limited. Therefore, there is no unified standard. Lan etc [9]. The study found that patients with early active chemotherapy can survive longer, and local radiotherapy and surgery can only relieve symptoms. New research suggests that hematopoietic stem cell transplantation is a new promising therapeutic direction. Tan, etc [10] have reported a patient with primary GS who recovered successfully by allogeneic stem cell transplantation after receiving systemic chemotherapy. At present, stem cell transplantation is probably the most effective treatment for GS.

For diagnosing such stomach tubercle patients, both gastroenterology doctors and pathology doctors should be sensitive enough to confirm whether it is GS or not, which is very important for further treatment.

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