CASE REPORT

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Gastrointestinal Stromal Tumor of the Duodenum in a 13-Year-Old Girl: A Rare Case Report

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ABSTRACT

Introduction: One of the main causes of death for kids and teenagers is a tumor which exceeds 200.000 in all of the world. One of the tumors is Gastro Intestinal Stromal Tumor (GIST). GIST is a mesenchymal gastrointestinal tumor with a focus on the Interstitial Cells of Cajal (ICC). Not more than 100 cases of pediatric GIST and less than 5% of childhood tumors are seen in the Gastrointestinal (GI) Tract. Due to their rarity, the current literature consists only of reviews of small case series, less than a third of all cases reported in the literature.

Case Description: A thirteen-year-old girl presented to the emergency room with nausea, dizziness, and syncope a day before. She had the same complaint 2 months before and was diagnosed with iron deficiency anemia but it got worse when she came to emergency in the next two months. She got some tests and found a tubulovillous adenoma suspected as GIST in duodenal. This girl was unable to survive in the intensive unit after the surgical resection due to the massive bleeding in her stomach.

Conclusion: We reported a rare case of Gastrointestinal Stromal Tumor (GIST) of the duodenum found in a girl.

Keywords: Gastrointestinal Stromal Tumor, Anemia, Tubuluvillous Adenoma.
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INTRODUCTION

Tumors are one of the leading causes of death among children and teenagers, with the incidence steadily increasing since 1975. Worldwide, the number of childhood tumor cases exceeds 200,000, with over 80% occurring in developed countries[1]. One such tumor is the Gastrointestinal Stromal Tumor (GIST). GIST is classified as a mesenchymal tumor of the gastrointestinal tract, originating from the Interstitial Cells of Cajal (ICC), which are believed to function as the pacemakers of the gastrointestinal tract[2]. While GISTs can develop anywhere in the GI tract, not all are malignant, and more than half originate in the stomach. GIST is the most common mesenchymal tumor in the adult gastrointestinal tract, but the situation is different in children under 18 years old, where fewer than 100 pediatric cases have been reported[3].

Less than 5% of childhood tumors occur in the gastrointestinal (GI) tract. Due to their rarity, the current literature consists mainly of small case series reviews[4], and only about a third of all cases have been documented in the literature[5]. The purpose of this paper is to report a GIST case encountered in our hospital.

CASE DESCRIPTION

A 13-year-old girl presented to the emergency department with complaints of nausea and dizziness that began the previous day. She had experienced a syncopal episode while at school. Two months earlier, she had a similar episode and was diagnosed with iron deficiency anemia. According to her medical records, she had previously received a blood transfusion due to dyspnea and retraction and had been prescribed iron supplements. She had not sought medical care at a health center before because she lived in a dormitory.

On physical examination, her blood pressure was 103/85 mmHg, her heart rate was 126 beats per minute, her oxygen saturation was 100% on room air, and her temperature was 37.6°C. She appeared pale upon arrival at the emergency department. She reported abdominal pain in the epigastric and right hypochondriac regions. A 6-7 cm mass was palpable in her abdomen near the umbilicus.

A complete blood count (CBC) test was performed upon her arrival, revealing some abnormalities.

Due to her anemia, a blood smear test was performed. The results indicated normocytic normochromic anemia, consistent with chronic anemia. In addition to the blood tests, she underwent an abdominal ultrasound, which revealed a 7.09 cm x 7.07 cm gastric tumor and mild splenomegaly. A stool test was also conducted to check for the presence of blood.

Her mother brought the results of her last blood test, which had been conducted two months earlier. She was retested upon arrival at the emergency department.

After all the tests, she was administered 2200 cc of Ringer's Lactate via intravenous line over the course of one day. She was also given 500 mg of metamizole for dizziness and stomach pain. A plan was made to provide one unit of packed red cells (PRC)

Table 1. Complete Blood Count

Laboratory	Result	Ref. Range
Complete Blood Count		
Hb	6,5	10-16 (g/dL)
Leucocyte	8.100	5.000 – 10.000 (mm³)
Eosinophils	-	1 – 3 %
Basophils	-	0 – 1%
Stabs neutrophils	-	2 – 6%
Segmented neutrophils	67	36 – 65 mg/dL
Lymphocytes	24	20 – 35%
Monocyte	9	2 – 6%
Erythrocyte	2.19	4 – 5m/mm³
Hematocrit	18.5	38 – 47%
MCV	83.0	80 – 100 fL
MCH	28.3	27 – 34 pgw
MCHC	34.1	32 – 36 g/dL
RDW – CV	14.5	11 – 16 %
RDW - SD	43.9	35 – 56 fL
Thrombocyte	207.000	150.000 – 500.000/mm³

Table 2. Stool Test

Laboratory	Result	Ref. Range
Stool Test		
Consistency	Soft	-
Color	Dark	-
Mucous	Negative	(-) Negative
Blood	Negative	(-) Negative
Leucocytes	(2 – 4)	(-) Negative
Erythrocyte	(0 – 1)	(-) Negative
Amoeba	Negative	(-) Negative
Eggs	Negative	(-) Negative
Digestion	Remaining meat	-
Others	Amylum	-

Table 3. Complete Blood Count, Iron Status, Immunohematology 2 Months Before

Laboratory	Result	Ref. Range
Complete Blood Count		
Hb	7	10-16 (g/dL)
Leucocyte	16.300	5.000 – 10.000 (mm³)
Eosinophils	-	1 – 3 %
Basophils	-	0 – 1%
Stabs neutrophils	-	2 – 6%
Segmented neutrophils	78	36 – 65 mg/dL
Lymphocytes	15	20 – 35%
Monocyte	7	2 – 6%
Erythrocyte	2.47	4 – 5m/mm³
Hematocrit	18.5	38 – 47%
Thrombocyte	207.000	150.000 – 500.000/mm³
Iron Status		
Serum Iron	30	49-151ug/dL
TIBC	283	250 – 350ug/dL
Transferrin saturation	11	16 – 45%
Immunohematology		
Coomb Test	Negative	Negative

daily for blood transfusion, along with 5 mg of dexamethasone as premedication.

While receiving the blood transfusion, she was referred to a larger hospital due to the tumor in her stomach. After being transferred, she underwent an abdominal CT scan, which revealed an intraperitoneal mass adhering to the minor gastric curvature. The mass was suspected to be either a gastrointestinal stromal tumor (GIST) or a mesenteric cyst.

She was scheduled for a duodenal biopsy to confirm the tumor type. The biopsy revealed a tubulovillous adenoma with mild dysplasia. A few days after the biopsy, she underwent open surgical resection. Unfortunately, she did not survive post-operatively in the intensive care unit due to massive abdominal bleeding.

DISCUSSION

Gastrointestinal Stromal Tumors (GIST) are rare tumors of the gastrointestinal tract and are not always cancerous[3]. Cancer of the gastrointestinal (GI) tract is uncommon in children, with less than 5% of childhood tumors occurring in the GI tract[4]. Only up to 2% of GIST cases are diagnosed in children and adolescents, and they typically present as sporadic tumors[5]. A systematic review of published studies indicates that GIST affects girls more frequently than boys, as seen in this case. There is no significant difference in the median age of diagnosis between female and male patients, which is 14 years for females and 15 years for males[6]. In this case, the patient, a 13-year-old girl, was diagnosed with GIST, which aligns with the findings of the published study.

In 10–30% of patients, GIST is asymptomatic. However, there were notable clinical manifestations in this case. Acute or chronic GI bleeding is the most common clinical presentation, typically manifesting as general weakness and/or syncope[7]. While this patient did not have GI bleeding, she did report a syncopal episode the day before. Other nonspecific symptoms and signs include loss of appetite, abdominal pain or distension, nausea, vomiting, constipation, epigastric discomfort, diarrhea, intestinal obstruction, or the presence of an abdominal mass. In this case, the patient

Figure 1. The abdominal CT scan shows an intraperitoneal mass that sticks to minor gastric curvature.

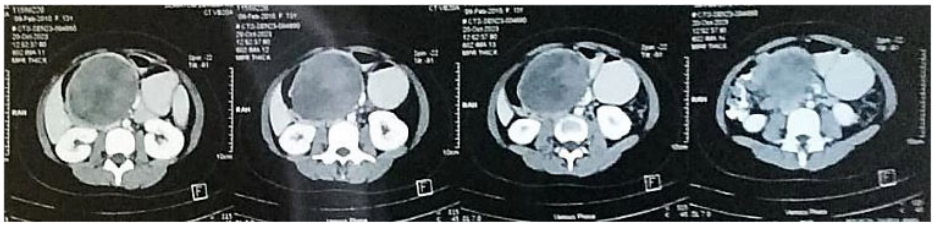

Figure 2. The abdominal CT scan shows a 7,09 cm x 7, 07 cm mass in her duodenal.

exhibited several clinical manifestations, such as abdominal pain and a palpable abdominal mass.

GISTs are typically asymptomatic until they reach a size of about 6 cm. These larger tumors are often vascular and present with significant symptoms[8]. In this case, although she was diagnosed with iron deficiency anemia two months prior, there was no palpable abdominal mass at that time.

Seven cases of GIST with moderate to mild anemia have been reported in both pediatric and adult patients, with hemoglobin levels ranging from 8 to 10.2 mg/dL[8,11]. In this case, the patient presented with a hemoglobin level of 6.5, which is lower than the range reported in previous cases.

In this case, the patient's hemoglobin level dropped by 0.5 mg/dL despite iron supplementation. She had been prescribed iron supplements after being diagnosed with iron deficiency anemia. This is similar to other reported cases, where patients experienced significant hemoglobin drops, with an average decrease of 3 g/dL, after pharmacological treatment over varying periods of time—ranging from two days to three and eight months[7,8,10-14]. In our patient, the drop occurred over two months. This suggests that GIST should be considered as a differential diagnosis in cases of moderate anemia with worsening hemoglobin levels and symptomatology. Any child presenting with persistent or acute, severe, refractory idiopathic anemia, with or without GI symptoms, should be evaluated for GIST[7].

A duodenal incisional biopsy is the preferred technique for confirming a primary diagnosis. In this case, the biopsy revealed a tubulovillous adenoma with mild dysplasia. Adenomas are more commonly found in the colon and are rare in the small intestine. Duodenal tubulovillous adenomas are extremely uncommon, accounting for only about 1% of all duodenal tumors[15]. These adenomas carry a 20% to 25% risk of progressing to malignancy[16]. Diagnosing small bowel tumors is traditionally challenging, but imaging techniques like abdominal CT scans can assist in diagnosis[17]. In this case, an abdominal CT scan confirmed the presence of GIST.

The preferred treatment for GIST is surgery without lymph node removal[5,8]. Small gastric GISTs (<5 cm) can be managed laparoscopically, but larger tumors typically require open surgery to avoid rupture and minimize the risk of peritoneal dissemination[8]. In this case, the abdominal CT scan revealed a tumor measuring 7.09 cm x 7.07 cm, categorizing it as a large tumor. The patient underwent open surgery but unfortunately did not survive the postoperative period.

CONCLUSION

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors of the gastrointestinal tract that rarely occur in pediatric patients. Due to their rarity, there are only a few reviews on pediatric GIST cases. Many patients with GIST show no clinical symptoms until the tumor reaches 6 cm in size, which can lead to late diagnosis. Biopsy is crucial for establishing a definitive diagnosis. Surgical procedures without lymph node removal remain the standard treatment. Although many cases are successfully treated with surgery, follow-up is essential due to the risk of recurrence.

DISCLOSURES

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None.

Conflict of Interest

The authors declare that there are no conflicts of interest.

Author Contribution

YP: concepts, literature search, manuscript preparation, manuscript editing. SM: design, definition of intellectual content, literature search, manuscripts review.

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