Gastric adenocarcinoma in childhood: a case report and a literature review

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ABSTRACT
Gastric tumors, especially gastric adenocarcinomas, are rare in childhood and adolescence, as a result of which there is limited information. Therefore, management is typically extrapolated from adult patients.

We report the case of a 10-year-old girl referred to our institution with systemic symptoms and pyloric syndrome. An infiltrating antro-pyloric lesion without evidence of metastasis was found. Histopathological analysis confirmed the presence of an undifferentiated diffuse gastric adenocarcinoma with signet ring cells. Diagnostic laparoscopy was carried out, which demonstrated signs of peritoneal carcinomatosis, so palliative chemotherapy was proposed.

KEY WORDS: Gastric cancer; Adenocarcinoma; Gastric neoplasia; Child.

INTRODUCTION
Gastric cancer is a common pathology in the adult population. According to Globocan statistics, it is the fifth most frequent cancer and the third cause of death globally. However, it is rare in the pediatric population, where gastrointestinal neoplasia represents 5% of all cancers, with gastric adenocarcinoma accounting for 0.05%. Most gastric adenocarcinomas are diffuse, with signet ring cells associated with metastasis or carcinomatosis, which makes prognosis worse. Data on pediatric patients are limited.

RISK FACTORS FOR GASTRIC ADENOCARCINOMA INCLUDE DIET, LIFESTYLE, AND HELICOBACTER PYLORI INFECTION. IN ADDITION, GENETIC FACTORS PLAY A ROLE IN 10% OF CASES, AND PROBABLY MORE IN THE_pediatric population. Gastric cancer can be either intestinal or diffuse, the latter being more common in younger patients. Diffuse gastric adenocarcinoma has been demonstrated to be caused by a molecular abnormality in e-cadherin cell adhesion protein (CDH1), and typically occurs in families with a hereditary diffuse gastric cancer dominant autosomal trait. CDH1 gene is involved in 25-36% of cases, most of which have signet ring cells associated with intestinal involvement. In intestinal gastric cancer, progression from chronic gastritis to atrophic chronic gastritis, intestinal metaplasia, dysplasia, and eventually adenocarcinoma has been described. Symptoms are unspecific. Management varies according to disease stages. In our environment, most cases are advanced stages in adult patients.

CASE PRESENTATION

10-year-old girl referred to our institution after 2 months of non-irrigated abdominal pain at the epigastrium, associated with episodes of postprandial vomiting and a 7 kg weight loss. She had no pathological or surgical history, but she did have family history of colon cancer – a paternal
Aunt. She showed a nice condition at physical exploration, without lymph node enlargement. The abdomen was soft, with a hard, non-mobile epigastric mass adhered to the deep planes.

Abdominal ultrasonography revealed gastric wall thickening. Upper GI endoscopy showed signs of gastric retention with gastric wall rigidity and infiltrating lesion of the gastric body, as well as a gastric antrum with mucosal enhancement and reduced luminal caliber. Multiple biopsies of the lumen were carried out (Fig. 1). An advanced endoscopic nasogastric tube was placed for enteral feeding purposes. Thoracoabdominal CT-scan found concentric wall thickening at the antro-pyloric region, with perigastric lymph nodes both at the greater and the lesser curvature, the gastrohepatic ligament, and left para-aortic lymph nodes of up to 6 mm, without evidence of hepatic or pulmonary metastases (Fig. 2). The histopathological study demonstrated an undifferentiated diffuse gastric adenocarcinoma, with signet ring cells and cytokeratin, leukocyte common antigen, e. cadherin, MSH6, MLH1, MSH2, and PMS2 positive immunohistochemistry (Fig. 3). Tumor markers were measured, with 1.37 ng/ml (< 2.5 ng/ml) carcinoembryonic antigen and 146 U/ml (0-35 U/ml) carbohydrate antigen 125 levels. Diagnostic laparoscopy was carried out for abdominal compromise assessment and surgical planning purposes. It showed lesions suggestive of peritoneal seeding in both hemidiaphragms, where biopsies were performed, which found serosanguinous peritoneal fluid. A 250ml cell block sample was taken. The stomach had diffuse body and antrum thickening. Mobilization could not be achieved as a result of how strongly the antro-pyloric region was adhered to the pancreatic body. No further lesions were observed. Peritoneal fluid histopathological study demonstrated isolated mesothelial cells and cell population with mild atypia. The biopsy of the right hemidiaphragm showed no signs of malignity, while the biopsy of the left hemidiaphragm revealed metastatic compromise as a result of carcinoma. The patient was classified as stage IV, so cisplatin and 5-fluorouracil chemotherapy was initiated with palliative purposes. 4 months following diagnosis and chemotherapy initiation, an abdominal CT-scan was conducted, which showed disease progression with ascites, peritoneal thickening, and increased gastric wall thickening. The patient died 7 months following diagnosis.

Figure 1. Endoscopic finding demonstrating an infiltrating lesion at the antro-pyloric region.
DISCUSSION

Various elements are to be considered in the development of gastric cancer, including environmental factors such as diet, lifestyle, and Helicobacter pylori infection. Even though most gastric cancers are sporadic in adults, familial factors play a role in approximately 10% of the cases. Gastric cancer represents 1-3% of the global burden, and it has been described in Lynch’s syndrome, familial adenomatous polyposis, Li Fraumeni syndrome, Peutz Jeghers syndrome, and juvenile polyposis.

According to histological appearance, gastric cancer can be either intestinal or diffuse, and according to differentiation degree, it can be classified as well differentiated, moderately differentiated, and poorly differentiated. The signet ring cell feature is associated with a worse prognosis.

Gastric cancer clinical manifestations can be unspecific in early cancers and highly remarkable in advanced stages, with dyspepsia, epigastric pain, nausea, anorexia, weight loss, abdominal mass, lymphadenopathy, and digestive bleeding. Endoscopy with systematic biopsies of suspicious lesions is the diagnostic procedure of choice, with a 98% sensitivity and a 70% specificity. The stage is determined according to the TNM classification. Tumor markers such as carcinoembryonic antigen (CEA), carbohydrate antigen 125 (CA 125), and carbohydrate antigen 19-9 (CA 19-9) can be high, but they are little sensitive and specific. Diagnostic laparoscopy, in spite of being invasive, allows the intra-abdominal surface to be visualized, and samples to be taken for peritoneal cytology, which can change patient management in many cases.

Tumors with local and regional disease are potentially resectable (stages I-III), whereas tumors with advanced disease (stage IV) are not (Table 1). Gastric cancer treatment includes endoscopic resection, neoadjuvant therapies, and surgery. Total or subtotal gastrectomy is required, according to tumor location. Survival in the pediatric cases reported in the literature ranges from 5 months to 8 years following diagnosis. The study by Vivek Subbiah et al., carried out in 292,621 cancer patients, identified 5 pediatric patients with gastric cancer, which accounts for 0.0017% of all cases, 0.11% of all gastric cancer patients, and 0.08% of all cancer patients under 18 years of age. Median age was

Figure 2. Contrast abdominal CT-scan. The white arrows show gastric wall thickening.

Figure 3. 40X light microscopy. Hematoxylin and eosin staining reveal the presence of signet ring cells.
Adenocarcinoma is a rare neoplasia in childhood. There are no management algorithms in pediatric patients, which means they have to be extrapolated from those of adults. In addition, pediatric surgeons and oncologists usually lack the experience required to deal with it. Staging and therapy in children is based on oncological experience in adults.

Even though our patient did not undergo genetic tests, identifying familial diffuse gastric cancer will allow these tests to be carried out in patient relatives. CDH1 mutation patients should be approached using screening endoscopy and/or prophylactic gastrectomy, whereas women should undergo breast surveillance as a result of breast cancer risks.

**CONCLUSIONS**

Adenocarcinoma is a rare neoplasia in childhood. There are no management algorithms in pediatric patients, which means they have to be extrapolated from those of adults. In addition, pediatric surgeons and oncologists usually lack the experience required to deal with it.

**REFERENCES**


