CASE REPORT

Linitis plastica in a 14-year-old boy: A rare form of gastric cancer encountered in southern Ethiopia

Joseph Bedore[1], Kibruyisfa Desalegn[1], Adnan Alseidi[2]

[1]Hawassa University College of Medicine and Health Sciences, Hawassa, Ethiopia
[2]Division of Surgical Oncology, Department of Surgery, University of California San Francisco, San Francisco, CA, USA

Correspondence: Dr Joseph Bedore (josgive@yahoo.com)

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Abstract

Linitis plastica, a morphological variant of diffuse infiltrative adenocarcinoma of the stomach, is an extremely rare cancer in children. Its clinical presentation, treatment, and outcomes are scarcely described in the literature. We report a case of gastric cancer, specifically linitis plastica, in a 14-year-old boy who presented with obstructive jaundice. Ultrasonography revealed free peritoneal fluid, circumferential gastric wall thickening, and grossly dilated intrahepatic and extrahepatic biliary ducts. Intraoperatively, massive ascites, peritoneal carcinomatosis, and diffuse infiltration of the stomach were observed, rendering the cancer unresectable. Biopsies were subsequently taken. The patient died 2 months postoperatively.

Keywords: gastric adenocarcinoma, linitis plastica, peritoneal carcinomatosis, obstructive jaundice, paediatric surgery, surgical oncology, Ethiopia

Introduction

Gastric adenocarcinoma is an extremely rare malignancy. It has a grave prognosis, notwithstanding attempts at treatment with various modalities.[1] Its presentation is nonspecific, and few cases of paediatric linitis plastica have been reported, each with unique clinical presentations. While gastric cancer can cause biliary obstruction—frequently via metastatic lymphadenopathy at the hepatoduodenal ligament[2]—this is an uncommon presentation of the disease.

Case presentation

A 14-year-old boy was referred to our tertiary referral centre in southern Ethiopia with chief complaints of right-upper-quadrant and epigastric pain, along with occasional nonbilious vomiting over a 4-month period. During this time, he experienced progressive and significant (albeit unquantified) weight loss. Two months after the onset of these symptoms, he developed yellowish discolouration in his eyes, cola-coloured urine, and clay-coloured stool. He showed no signs of upper or lower gastrointestinal bleeding and had no prior history of dyspepsia. He had been fully vaccinated according to the World Health Organization's Expanded Programme on Immunization.[3] His family history was negative for chronic medical diseases, gastrointestinal malignancies, or peptic ulcer disease. Two weeks before his admission to our hospital, he was admitted to a district hospital with high-grade intermittent fever. Due to a lack of clinical improvement, he was referred to our hospital for further investigation.

Upon physical examination at our centre, the patient was conscious, oriented, emaciated, and generally appeared unwell. His pulse was 114 beats per minute, with blood pressure normal for his age, a respiratory rate of 24 breaths per minute, and an oxygen saturation of 98% on room air. His axillary temperature was 37.5 °C. With a weight of 26.9 kg and height of 138 cm, he was underweight and stunted, both being below the third percentile for his age and sex. His conjunctivae were pale, and his sclerae were deeply icteric. Neither palpable pathological lymph nodes nor peripheral oedema were detected. Examinations of his respiratory and cardiovascular systems were normal. His abdomen was generally distended, with his liver and spleen ballotable below the costal margin. A digital rectal examination revealed clay-coloured stool with no blood detected on the examining finger. Given these findings and his severe malnutrition, he was admitted to our surgical service for suspected obstructive jaundice and ascending cholangitis.

Laboratory tests revealed a white blood cell count of 3215 cells/μL, with neutrophilia of 83.6% and a lymphocyte differential count of 5.64%. His haematocrit was 19.4%, and his platelet count was 602.7 × 10^3 cells/μL. The *Helicobacter pylori* test was positive, liver transaminases were within the normal range, and alkaline phosphatase was 1569 U/L. Total bilirubin was 16 mg/dL, and direct bilirubin was 5.97 mg/dL.

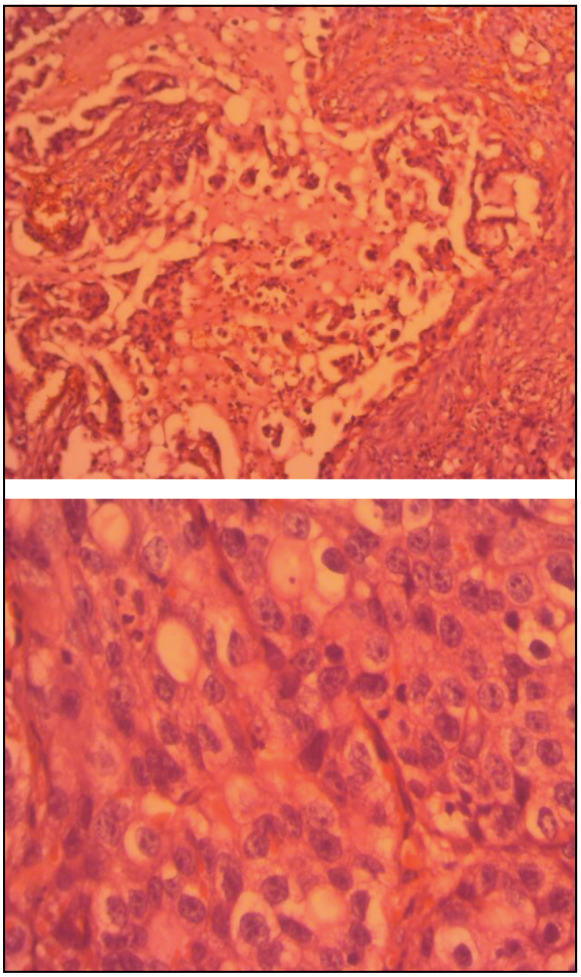

Figure. Surgical biopsy micrograph showing features of linitis plastica (diagnosed in a 14-year-old boy managed at a tertiary hospital in southern Ethiopia)

Hyperchromatic, pleomorphic, round-to-ovoid cells with prominent nucleoli and eosinophilic, granular, and vacuolated cytoplasm. The cells are arranged in solid sheets and nests, with some forming glandular structures. Areas of necrosis are visible, along with frequent mitotic figures. The desmoplastic stroma features mixed inflammatory cells and areas of haemorrhage.

Abdominal ultrasonography revealed free peritoneal fluid, a collapsed stomach with circumferential wall thickening of 13 mm, and heterogeneous thick omenta with small hypoechoic nodules. The gallbladder contained luminal sludge, and there was significant dilatation of the intrahepatic and extrahepatic biliary ducts. Abdominal computed tomography and endoscopy could not be performed owing to financial constraints. The patient received intravenous ceftriaxone, metronidazole, and vitamin K. Given his clinical deterioration, an exploratory laparotomy was deemed necessary.

Differential diagnosis

Hepatic jaundice is the most commonly encountered mechanistic category of jaundice in children.[4] The common causes of obstructive jaundice in children differ from those in adults.[5] Differential diagnoses in children include stone disease, congenital anomalies, parasitic infections, inflammatory conditions, and rare malignant diseases.[1],[5] Beyond causes of obstructive jaundice originating from the hepatobiliary system, there are rare causes arising from other structures in the vicinity of the liver and biliary tree, as observed in our patient. Fibrosing pancreatitis is among the rare causes of obstructive jaundice in children.[4] Lymphoma can also present with signs and symptoms of obstructive jaundice, though this is not its usual presentation in children.[6] These aetiologies are among the rare causes of obstructive jaundice beyond the neonatal period. Biliary atresia, inspissated bile syndrome, and choledochal cysts are causes of jaundice in the neonatal age group.[7] In rare instances, tuberculosis (TB) can cause obstructive jaundice due to pressure effects on the porta hepatis.[8] Tuberculosis can also cause biliary stricture and present with jaundice, although this is not commonly observed following infection by TB.[9]

Management

The patient received intravenous ceftriaxone, metronidazole, and vitamin K. Due to clinical deterioration, an explorative laparotomy was performed. The abdomen was accessed through a high midline incision, revealing 2 L of ascitic fluid, a diffusely thickened stomach, diffuse peritoneal carcinomatosis, and enlarged porta hepatis lymph nodes causing a mass effect on the biliary tree. No nodularity was observed on the liver surface. The ascitic fluid was aspirated, and biopsies were taken from the omentum and falciform ligament. Histopathology revealed metastatic, poorly differentiated signet ring cell adenocarcinoma originating from the stomach (Figure). Outcome and follow-up

After a 2-week stay at our institution, the patient was discharged with a follow-up appointment scheduled. At his first visit nearly a month later, he presented with generalized oedema, deeply icteric sclera, pallor, and severe emaciation. By his second visit, there was a significant clinical deterioration; he progressed to multiorgan failure and died after nearly 2 postoperative months.

Discussion

We present an exceedingly rare case of linitis plastica in a 14-year-old boy who presented with obstructive jaundice.

Linitis plastica, a diffuse and infiltrative gastric adenocarcinoma, typically involves the entire stomach, leading to transmural thickening and stiffness, often described as a 'leather bottle stomach'. The primary cancer cells are usually signet ring cells or scattered cells characteristic of a poorly differentiated adenocarcinoma.[10],[11] Given the rarity of gastric adenocarcinoma in children and its nonspecific clinical presentation, diagnosis is often delayed, leaving optimal treatment and survival data unclear. The most common initial symptom is abdominal pain, followed by nausea, vomiting, loss of appetite, and general fatigue.[12],[13] Early diagnosis is rare, and the presence of symptoms usually indicates advanced disease. However, to our knowledge, no study to date confirms that early diagnosis confers survival benefits in paediatric gastric cancer. Despite the common scenario

of diagnosing these patients at stage 4 with poorly differentiated histopathology, paediatric patients seem to have equivalent outcomes to adults.[14]

Our patient presented with stage 4 gastric cancer with obstructive jaundice due to the mass effect of metastatic porta hepatis lymph nodes. At the time of diagnosis, the tumour was deemed unresectable, and the patient did not receive any systemic or cancer-directed therapy during his disease course. He died from advanced gastric cancer 6 months after the onset of his symptoms. Of the other children reported with linitis plastica of the stomach, at least 1 patient had a tumour that was surgically resectable at the time of diagnosis. This patient underwent a subtotal gastrectomy followed by adjuvant chemoradiotherapy and was reported to be disease-free at 1 postoperative year.[1] Two other patients with unresectable disease had survival times of 6 and 2.5 months post-diagnosis, respectively.[4],[15] Owing to the rarity of paediatric linitis plastica, it is challenging to gather meaningful data about the relative prognosis and survival rates relative to similar tumours in adults.

Conclusions

Gastric adenocarcinoma is a rare malignancy in children, with the linitis plastica variant being exceedingly rare.

The establishment of an international tumour registry to collect such rare cases for further histopathologic, genetic, and molecular evaluation could enhance our understanding of the disease and potentially offer more therapeutic options for these patients.

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