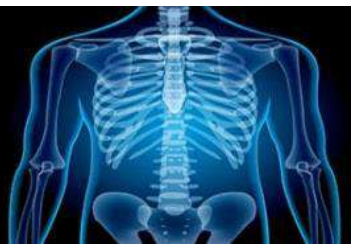
International Journal
of
Radiology and Diagnostic Imaging



E-ISSN: 2664-4444
P-ISSN: 2664-4436
www.radiologypaper.com
IJRDI 2022; 5(4): 86-89
Received: 05-08-2022
Accepted: 09-09-2022

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Menetrier's disease: A rare diagnosis established by ultrasonography and computed tomography

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DOI: http://dx.doi.org/10.33545/26644436.2022.v5.i4b.289

Abstract
Menetrier's disease is a rare condition of the stomach characterized by giant mucosal folds in the gastric fundus and body, diminished acid secretory capacity, and a protein-losing state with hypoalbuminemia. It is also called protein-losing hypertrophic gastropathy. It was first described by the French pathologist Pierre Menetrier in 1888. Menetrier's disease has also a recognized premalignant potential although the precise risk of progression to gastric cancer is not known. Several studies reported regression of disease after treatment with the monoclonal antibody against the EGFR receptor. But the only satisfactory treatment has historically been and remains the surgical intervention with total or partial gastrectomy.

A case report of A 10-year male presented to the emergency room with a history of progressively increasing abdominal pain on and off for 1 month with epigastric region fullness, nausea, and vomiting. Generalized edema was noted on examination. An abdominal ultrasound examination was performed, and the patient was found to have diffuse mucosal thickening of the gastric wall with prominent rugal folds and echogenic mucosal lining with generalized subcutaneous edema and mild ascites. The patient underwent surgery and the diagnosis was confirmed by the histopathological report.

Keywords: Menetrier's disease, protein-losing hypertrophic gastropathy, giant mucosal folds, premalignant potential, gastric cancer, and Monoclonal antibody against EGFR receptor

Introduction

Menetrier's disease is a rare condition of the stomach characterized by giant mucosal folds in the gastric fundus and body, diminished acid secretory capacity, and a protein-losing state with hypoalbuminemia [1]. It is also called protein-losing hypertrophic gastropathy. It was first described by the French pathologist Pierre Menetrier in 1888 [2]. Several studies reported regression of disease after treatment with the monoclonal antibody against the EGFR receptor [3]. Menetrier's disease has also a recognized premalignant potential although the precise risk of progression to gastric cancer is not known [4]. But the only satisfactory treatment has historically been and remains the surgical intervention with total or partial gastrectomy [5]. Menetrier's disease (MD) is a rare disease characterized by hypertrophic folds in the body of the stomach, foveolar hyperplasia, and hypoproteinemia due to the selective loss of serum proteins through the gastric mucosa [6]. MD usually occurs in children under 10 years of age, and boys are affected more often than girls [7]. This is an unusual case of pediatric MD without self-limitation needs specific treatment was also described in a previous study [8].

Case Report

A 10-year-old boy was admitted to the emergency department of Era Medical College and Hospital Lucknow with a history of on and off increasing abdominal pain for 1 month and abdominal fullness, nausea and vomiting. On examination, generalized subcutaneous edema was noted. Initial examination: temperature 37.8 °C, heart rate 98 beats/min, blood pressure 130/72 mmHg. Laboratory tests showed low levels of total protein (32.99 g/L, reference range: 60–80 g/L), albumin (24.82 g/L, reference range: 38–54 g/L) and globulin (14 g /l, reference range: 22-34 g/l). The hemoglobin level was normal, the reticulocytes were slightly upregulated and the percentage of eosinophils was increased (11%). Coagulation function and erythrocyte sedimentation rate were normal.

Abdominal ultrasound was performed at the Era Radiology Department by two experienced sonographers using a SAMSUNG HS 70A USG with a 7.5 MHz -11 MHz linear transducer (Samsung Electronics Pvt. Ltd., Seoul, South Korea). After suspicious findings on ultrasonography, contrast-enhanced CT was performed on a 384-slice dual-energy CT scanner (Somatom Force, Siemens Healthcare, Erlangen, Germany).

Sonographic localization of the stomach is typically performed with epigastric transducer placement with the patient in the supine or right lateral decubitus position; Begin by locating the gastroesophageal junction (GEJ) by placing the probe just to the left of the xiphisternum and rotating so that the probe points toward the right shoulder. GEJ will see deep into the left lobe of the liver below the diaphragm and deep into the heart above. To assess the rest of the stomach, locate the pylorus or first part of the duodenum in the right upper quadrant (RUQ). From there, move the probe to the left to assess the short axis of the stomach. The probe will need to be angled at the left costal margin to assess the body and fundus. To assess the long axis, hold the probe transversely over the epigastric region of the abdomen and fan the probe (cranial to caudal). Repeat this (from left to right) until you can see enough of the entire stomach.

Ultrasonography demonstrated diffuse mucosal thickening of the gastric wall with prominent rugal folds and an echogenic mucosal lining with generalized subcutaneous edema and mild ascites. There was a slight increase in the echogenicity of the liver. [Figure.1 and 2]. USG showing thickened gastric mucosa with prominent rugae. CT showed giant cerebriform enlargement of the rugal folds on the fundus and body of the stomach and minimal effusion in the pelvic cavity. [Figure. 3 and 4].

Discussion and Result

The diagnosis of Menetrier's disease was made during the diagnostic process and finally confirmed by histopathology. After informed consent, an endoscopic biopsy was performed. Gastrointestinal endoscopy revealed enlarged gastric folds, erythema, and hemorrhagic erosions covered with whitish mucus throughout the gastric body. Histological biopsies of the stomach showed foveolar hyperplasia with glandular atrophy and eosinophil infiltration in the patient's lamina propria. Histological examination (Fig. 5-8) showed diffuse foveolar hyperplasia with cystic dilatation of the glandular part of the gastric mucosa and the absence of a significant inflammatory infiltrate. Giant mucosal hypertrophy and hyperplasia consist mainly of gastric mucus cells, while parietal and chief cells are markedly reduced.

Although pediatric MD typically has an abrupt onset, is self-limited, and has an overall benign course that may resolve spontaneously within 2 to 10 weeks with supportive therapy alone [9] The child with MD described in this study was a boy less than 11 years of age. Studies suggest that pediatric patients with MD may have a variety of nonspecific symptoms. Common symptoms of pediatric MD include abdominal pain, nausea, frequent vomiting, diarrhea, loss of appetite, weight loss, and malnutrition [10]. Peripheral edema due to hypoalbuminemia is also often present in pediatric patients with MD [7, 8, 10]. In this report, the patient presented with symptoms of abdominal pain and vomiting, and eyelid edema. Radiological, endoscopic, and pathological findings further supported the diagnosis of MD. Finally, after 2 weeks of supportive treatment, the disease was cured and the boy was asymptomatic during a follow-up of 6 months.

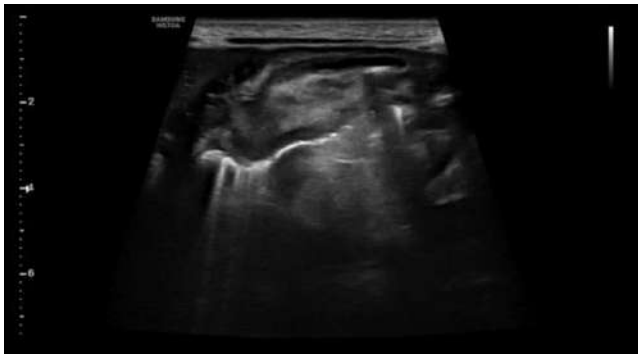

Fig 1: Grayscale ultrasound showed -thickened gastric mucosa with prominent rugae

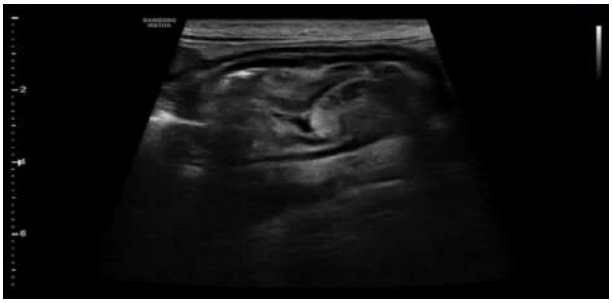

Fig 2: Grayscale ultrasound showed - diffuse mucosal thickening of gastric wall with prominent rugal folds and echogenic mucosal lining with generalized subcutaneous edema and mild ascites

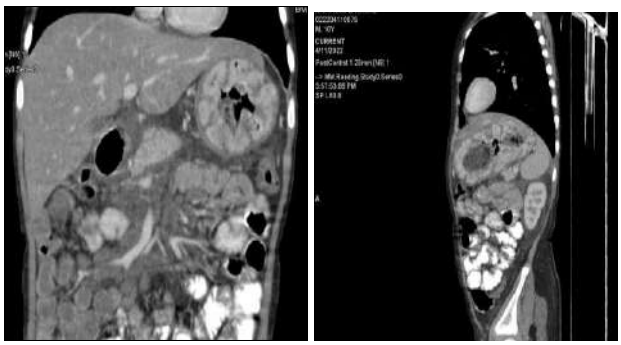

Fig 3: Abdominal computed tomography (CT) images obtained with intravenous Omnipaque showing diffuse gastric wall thickening, mucosal hyperenhancement, and submucosal edema

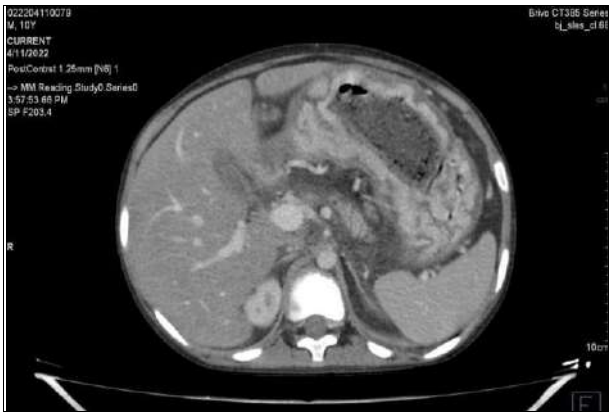

Fig 4: Abdominal computed tomography (CT) images obtained with intravenous Ominpaque showing giant cerebriform enlargement of rugal folds in the gastric fundus and body

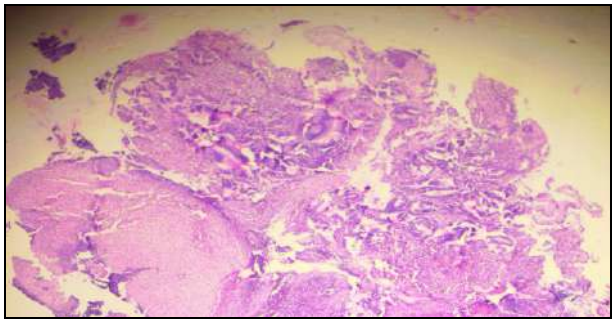

Fig 5: Microscopic features of the menetriers disease (hematoxylin and eosin stain) showing foveolar hyperplasia with glandular atrophy, infiltration of eosinophils, plasmocytes, and neutrophils in the lamina propria

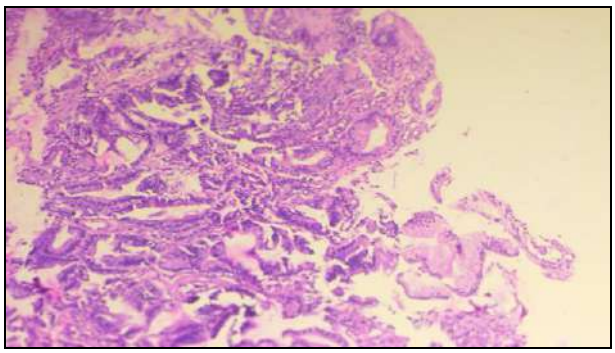

Fig 6: Microscopic features of the menetriers disease (hematoxylin and eosin stain) show elongated, tortuous, and cystically dilated foveolar glands, discontinuous atrophy of gastric glands, and significant reduction of parietal cells

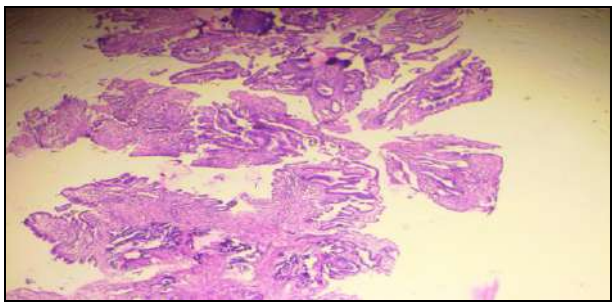

Fig 7: Microscopic features of the menetriers disease (hematoxylin and eosin stain) - showing Edematous and mildly inflamed lamina propria, may have increased intraepithelial lymphocytes

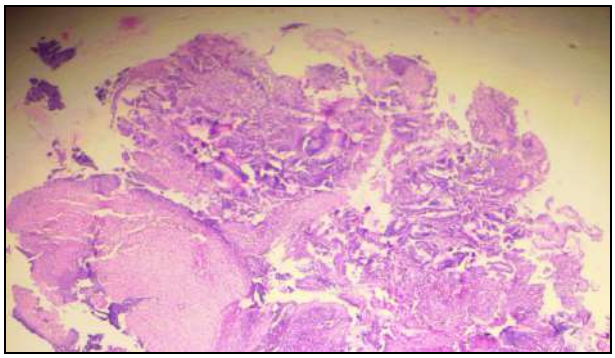

Fig 8: Microscopic features of the menetriers disease (hematoxylin and eosin stain) - showing marked foveolar hyperplasia, tortuous (corkscrew), and cystically dilated foveolar glands

Conclusion

In summary, in this study, we have described the clinical features of a pediatric case with MD from India. Due to nonspecific symptoms, gastrointestinal endoscopy with gastric tissue biopsy is required to establish the diagnosis of MD in children with unexplained hypoalbuminemia. Signs and symptoms are atypical. It should be differentiated from chronic hypertrophic gastritis, gastric polyps, gastric cancer, gastric stromal tumor, gastroptosis, and especially gastric lymphoma. Although the mucosa is greatly thickened, the lesions are limited to the mucosal layer only, so the morphology changes upon compression, the stomach wall is soft, and emptying is slowed. Radiological examinations (ultrasonography and computed tomography) are very useful for the diagnosis of MD, which helps in the early treatment of patients and the prevention of malignant transformation. Another gastroscopy may be performed when identification is difficult and confirmation of the diagnosis depends on pathological examination.

Acknowledgement
Not available

Conflict of Interest
Not available

Financial Support
Not available

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How to Cite This Article
K Sachin, G Ashkrit, P Vaibhav, Surbhi, K Zaara and D Vibhor. Menetrier's disease: A rare diagnosis established by ultrasonography and computed tomography. National Journal of Clinical Orthopaedics**.** International Journal of Radiology and Diagnostic Imaging 2022;5(4):86-89.