Ménétrier's disease in a child

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Summary: A case of Ménétrier's disease in a 3 year old child presenting with subtotal pyloric stenosis and fatal outcome due to postoperative complications is reported. It is emphasized that although radiographic and gastroscopic studies are helpful, a full-thickness mucosal biopsy is essential for the diagnosis of Ménétrier's disease.

Introduction

Ménétrier's disease (MD) or giant hypertrophy of the gastric rugae is an uncommon lesion first described in adults in 1888.¹ Besides more than 250 cases in adults, 31 cases of MD in children have also been reported since.² It was suggested that about 7% of all cases of MD occurred in children less than 15 years of age.³ The majority of children were aged 3–5 years old^{3,4} but the disease has occurred even in the neonatal period.^{5,6} As the course and prognosis of MD in children are different from those in adults, some authors regard it as a distinct disease which shares radiographic, endoscopic and biopsy findings with adult MD.⁷ MD in adults proves to be chronic and severe with a risk of developing gastric carcinoma.⁸ On the other hand, in most paediatric cases the disease is benign, self-limited and recedes within a few weeks.^{3,4,9}

In this report, we describe a child with MD who presented with severe gastric outlet obstruction requiring partial gastrectomy. In addition, the criteria for establishing the diagnosis of MD will be discussed.

Case report

A 3 year old boy was admitted to the local hospital with 15 days history of nausea and vomiting. An upper gastrointestinal barium study showed severe pyloric stenosis and laboratory tests revealed hypoproteinaemia with hypoalbuminaemia [total proteins 54 g/l (normal range: 60–80), albumin 26 g/l (38–46), globulins 28 g/l (24–32), ESR 7 mm/h, haemoglobin 11.0 g/l]. Explorative laparotomy

was performed 5 days later. No tumour infiltration or hypertrophic muscular tissue was found but giant hypertrophy of the antro-pyloric rugae was seen. Partial resection of hypertrophic mucosa was accomplished but 10 days after the operation radiology again demonstrated pyloric stenosis.

The child was referred to our Institute without a histological diagnosis. Total parenteral nutrition was initiated. On the next day, during vomiting, the child suddenly aspirated gastric content which was followed by generalized convulsions and, afterwards, by coma. After 10 days, the child showed some neurological signs of recovery. It was decided that the child should be operated upon. Subtotal pyloric stenosis with a 2 mm lumen was found. Partial gastrectomy was performed but the patient developed adult respiratory distress syndrome with oliguria and died one day after the operation. No autopsy was performed.

The microscopical findings in both specimens were characteristic for MD. The mucosa was markedly hypertrophic and hyperplastic (Figure 1) measuring 2 mm in thickness. Mucous metaplasia of the epithelium, marked foveolar hyperplasia, and elongated, tortuous glands were seen (Figure 2), with some dilated or even cystic glands, especially in the basal layer (Figure 3). In the lamina propria there was a severe mixed inflammatory cell infiltrate consisting of quite prominent eosinophils together with lymphocytes and plasma cells (Figure 3). The submucosa was oedematous, with dilated lymphatic channels and was infiltrated by inflammatory cells which had also extended into the muscularis propria.

Discussion

Giant rugal hypertrophy of the gastric mucosa associated with protein loss across an abnormal

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Figure 1 Full-thickness gastric biopsy demonstrating prominent thickened mucosa ($\times 63$).

Figure 2 Foveolar hyperplasia and elongated, tortuous gastric glands ($\times 240$).

gastric mucosa was originally described by Ménétrier in 1888 and has been well characterized since.³ Nausea, vomiting and oedema are usually leading clinical symptoms, while laboratory investigations always reveal hypoproteinaemia with hypoalbuminaemia.³ Gastric acid output studies, which in MD usually show hyposecretion,⁴ were not per-

Figure 3 Cystic dilatation of the basal gastric glands. An inflammatory infiltrate spreads from the lamina propria into the muscularis mucosae ($\times 160$).

formed in our patient due to the severe clinical course of disease. Upper gastrointestinal tract radiological and endoscopic studies have uniformly demonstrated large gastric rugae.^{3,4,9-11}

Although usually benign in children, the disease may follow a severe course, sometimes causing gastric outlet obstruction, as shown not only in the present case but in another reported case.⁶ Lethal outcome due to complications not directly related to the disease, as in our patient, has only been reported in two other cases.^{5,12} At times, partial or complete gastrectomy for correction of protein loss and hypoproteinaemia are required.^{5,6,10}

The aetiology of MD is still unknown although numerous aetiological factors have been postulated. It seems the possibility of allergy and/or viral infection is most likely in paediatric cases. Recent reports suggest a relationship between MD and cytomegalovirus (CMV) infection which has been proven in 11 paediatric patients in the world literature and, so, is probably not fortuitous.^{2,4} Since CMV infection is very common in humans (serological evidence of the previous infection is shown in 60% of population¹³) it is unlikely that CMV directly causes MD. However, CMV infection might be locally cytopathogenic, possibly allowing mucosal penetration of allergens which then stimulate a hypersensitivity reaction.⁴ In our case viral studies were not performed.

In the differential diagnosis of MD in children other conditions including eosinophilic gastroenteritis, hyperplastic hypersecretory gastropathy, Zollinger-Ellison syndrome, Crohn's disease and *Helicobacter pylori* gastritis, should be considered.^{4,11,14,15}

Some authors deem the diagnosis of MD can be made on clinical and radiological grounds only.⁴ It should be postulated here that radiological, endoscopic and clinical findings are of limited value since many cases diagnosed by radiologists and endoscopists as 'hypertrophic gastritis' may show

normal or atrophic mucosa on biopsy.¹⁶ Accordingly, histological examination of gastric mucosa is required for establishing the diagnosis of MD with certainty. A full-thickness biopsy of the gastric mucosa, obtained by a snare biopsy forceps, would provide a sufficient sample for analysis.¹⁵ The characteristic histological features are well-recognized and should not be missed or confused with other diseases which share similar clinical and/or radiologic features with MD.

Review of 31 previously reported paediatric patients revealed the diagnosis of MD was made without biopsy in 1/3 of cases.^{2,4} Critical evaluation of cases published before 1982 proved only two cases had qualified fully while the remaining lacked

pathological verification and were more difficult to evaluate.⁶ Therefore, we support the opinion that the term MD has been used too loosely and the diagnosis has often been made without using sufficiently rigorous criteria.⁶

On the basis of radiographic, gastroscopic and laboratory studies and histological examination, we believe the present case fulfils the rigorous criteria for MD, and serves to emphasize that a full-thickness gastric mucosal biopsy is essential for establishing the diagnosis of MD with certainty, while radiographic, gastroscopic and laboratory studies remain helpful but complementary methods only.

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