TRIGGER OF MÉNÉTRIER'S DISEASE: THREE CASES IN INFANCY

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Backgrounds

Ménétrier's disease is a rare, protein-losing gastroenteropathy, characterized by giant enlargement of gastric mucosal folds.
It typically presents with abdominal pain, vomiting and generalized edema secondary to hypoalbuminemia.
Esophagogastroduodenoscopy (EGDS) shows giant folds in the gastric body and fundus. Histopathology of the gastric mucosa typically reveals foveolar hyperplasia and decreased or absent oxyntic glands. Treatment is symptomatic as causal factors are not defined properly. Ménétrier's disease is extremely rare in children. Compared to the adult variant, Paediatric Ménétrier's Disease (PMD) usually has an acute, benign and self-limited course. The pathophysiology remains largely unknown, but the concomitant infection with Cytomegalovirus (CMV), found in 70% of pediatric cases, suggests the viral infection as possible trigger [(a)].

Aims

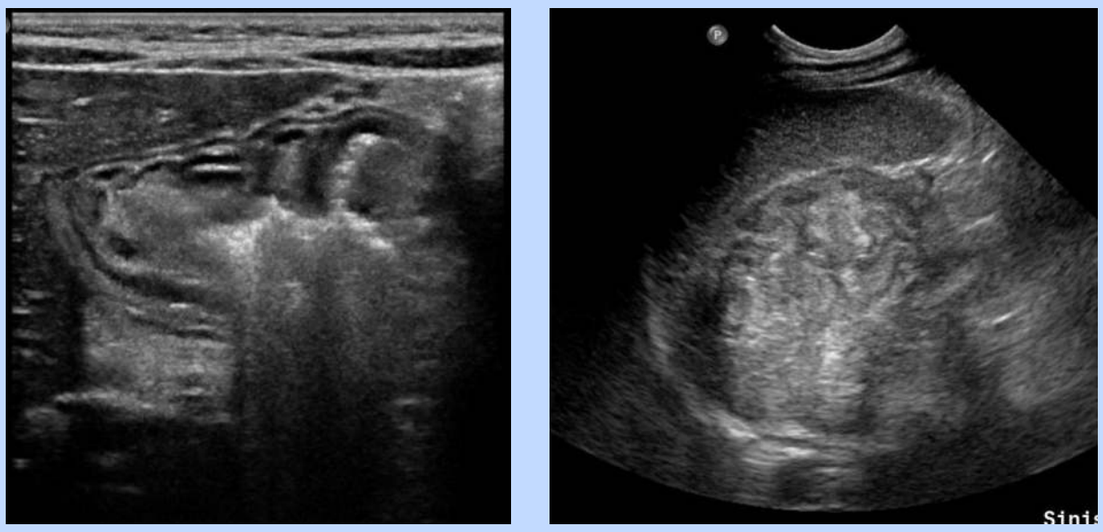
- To describe three children with classical presentation of PMD and positive detection of Cytomegalovirus;
- To focus on the specific investigations (CMV detection and EGDS) facing children with vomiting and diffuse edema.

Methods

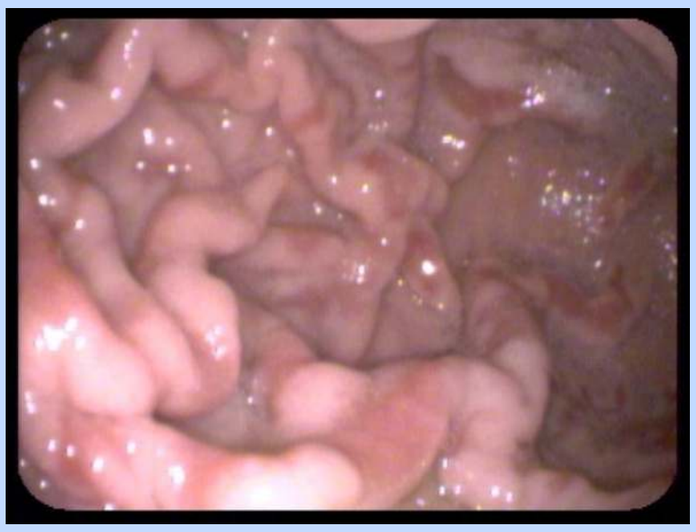
Medical records of three patients with PMD diagnosed at Anna Meyer Children's Hospital have been revised.

Results

- **Epidemiological data**: 3 males, aged 2 months to 3 years, previously healthy.
- **Clinical features**: protracted vomiting, periorbital and generalized edema present in all patients. The toddler showed also bloody diarrhea, dehydration and weight loss.
- **Laboratory data**: Severe hypoproteinemia and hypoalbuminemia in all cases, with no evidence of proteinuria or liver dysfunction. Coeliac disease was excluded.
- **CMV detection:**
  - CMV IgM and IgG antibody tests positive, with low IgG avidity.
  - CMV DNA detected by PCR in blood and urine samples.
- **Abdominal ultrasound** (US): bulging gastric folds in 1 patient; abdominal, pleural and pericardial effusions in another patient.
- **EGDS**: erythematosus gastric mucosa in 3 cases and hypertrophic gastric folds in the body and fundus in 2 of them.
- **Histopathological features**: foveolar hyperplasia and dilated gastric glands in all samples; CMV detected on biopsies of 2 patients, with CMV inclusion bodies or with viral DNA detection by PCR.
- **Treatment:** Intravenous albumin transfusions and diuretics administered in all patients, antivirals in 2 of them.
- **Outcome:** All patients had complete recovery.



I) Gastric US images showing bulging gastric folds



II) EGD image showing hypertrophic gastric folds and erythematosus gastric mucosa

Case	Age	Sex	Clinical presentation	Proteinemia (g/dL)	CMV detection	Radiology	Endoscopy	Histology	Management/ Outcome
1	3y	M	Protracted vomiting, periorbital and generalized edema	2.9	Positive serology	Abdominal, pleural and pericardial effusions	hyperthopic gastric folds, hyperaemic mucosa, erosions	Foveolar hyperplasia, CMV inclusion bodies	Supportive therapy. Recovery
2	2y	M	Protracted vomiting, drowsiness, asthenia, peripheral edema	2.9	Positive serology, CMV DNA by PCR	Bulging gastric folds	hypertrophic gastric folds, erythematosus gastric mucosa	Foveolar hyperplasia, dilated gastric glands, CMV DNA by PCR	Supportive and antiviral therapy. Recovery
3	2m	M	Protracted vomiting, bloody diarrhea, dehydration, weight loss	4.1	Positive serology, CMV DNA by PCR	Not performed	erythematosus gastric mucosa	Foveolar hyperplasia, dilated gastric glands	Supportive and antiviral therapy. Recovery

Conclusions

- PMD is very rare in children, with a benign and self-limiting course.
- Vomiting, hypoproteinemia with generalized edema, without renal and liver dysfunction, lead to protein-losing gastroenteropathy, such as PMD.
- CMV infection, as a possible trigger of Ménétrier's disease, needs to be investigated with specific serology and viral DNA detection by PCR on blood and urine samples.
- Abdominal ultrasound (hypertrophic gastric mucosa) may be helpful in the diagnosis.
- We recommend pediatricians to detect CMV infection and to perform gastroscopic examination with biopsies, facing children with vomiting and diffuse edema without renal and liver dysfunction.

References

a) Occena RO, Taylor SF, Robinson CC, Sokol RJ. Association of cytomegalovirus with Ménétrier's disease in childhood: report of two new cases with a review of literature. J Pediatr Gastroenterol Nutr 1993;17:217-24.