Medical cases in Paediatrics

Dr François Leung

Paediatrician

Medical Update Group

University of Mauritius

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Case No. 1:
T, 12½ m, female

- Diarrhea since 1 week
- Vomiting
- No fever
- Dehydration
- Abdomen: soft, distended++
- Pedal edema R>L
- Slight pubic edema
- W: 7.6 kg. L: 70 cm. Hc: 44 cm.

Case No. 1:
T, 12½ m, female

- Seen 3 days later
- Condition not better ... worse
- Weight 6.7 kg (Initially 7.6 kg)
- Stools: soft whitish, then liquid++.
- No vomiting
- Poor feeding
- Dehydration
- Pedal and pubic edema, ascites
- Abdominal girth: 44 cm.

Clinical Analysis

- Acute gastro-enteritis

- Unsual Features
  - Edema
  - Ascites

  → RVF ?

  → Portal HP ?

  → → → Hypoproteinemia / Hypoalbuminemia

Possible causes

Loss of proteins

- Urinary System
- Digestive System

  Inflammatory exsudatiion

  Increased mucosal permeability

  Intest. Loss of lymph. fluid

Insufficient intake/synthesis

- Malnutrition
- Maldigestion
- Malabsorption
- Impaired protein synthesis

Loss of proteins + Insufficient intake/synthesis

29.7.17: D1

- Proteinuria: Negative.
- Hb: 13.1. GB: 13 100. (Lc 60%) Plt: 408 000.
- Urea: 4.2. Electrolytes: 149/4.7/124.
- TG: 4.1 (↑). Chol T: 3.2 (↓).
- HDL: 0.8 (↓). LDL: 0.5 (↓).
- Prot: 37 (↓). Alb: 19 (↓). Glb: 18 (↓).
- Amylase: 77.
- CRP: neg.
- Stools: Culture neg, Rota/Adenovirus neg
- No parasite, no amoeba, no giardia/lamblia

Other tests:

- Chest Xray: Normal
- Echography: Gross free ascites

Treatment

- Albumine 10 g / day (x3)
- IV fluid
- Low-lactose milk: Adialac
- Prebiotics: Ultralevure
- Metronidazole
- Pancreatic enzymes: Créon
- Gluten-free diet
- Multivitamins
- → General condition improved grossly: ↑ Weight

3.8.17: D6

- Persistent diarrhea
- Massive liquid diarrhea on D6
- New loss of weight / dehydration. (W: 6.6 kg).
- Hb 12.3. WBC 12 000 (Lc58%). Plt 479 000.
- Urée 4.5. Elect 153/4.3/129. CRP neg.
- Prot 49. Alb 33. Glb 16.
- CMV Antibodies IgG & IgM: neg.
- Helicobacter pylori: negative.

→ → → Adialac replaced by Isomil

6.8.17: D9

- Diarrhea
- New massive diarrhea on D9 (W: 6.7 kg)

- Hb 11.6
- WBC 25 700 (N 58%, Lc 38%)
- Plt 316 000
- CRP <6 mg/l
- Urea 3.0 mmol/l. Electrolytes 136 / 4.9 / 107
- Prot 52 g/l Alb 34 g/l Glb 18 g/l

Observations

- 26.7.17. 7.6-70-44. Referred for admission
- 29.7.17. D1: 6.7 kg. (44) Adialac
- 1.8.17. D4: 7.7 kg. (44) Adialac
- 2.8.17. D5: 7.0 kg. (44) Adialac
- 3.8.17. D6: 6.6 kg. (42) Diarrhea+++
- 4.8.17. D7: 6.6 kg. (41) Isomil.
- 6.8.17. D9: 6.7 kg. (41) Diarrhea+++

Solution???

Persistent malabsorption

- Ac IgA antitransglutaminases (tTG IgA) >129 (n<7).
- Ac IgG antitransglutaminases (tTG IgG) <5 (N<7).
- Ac IgA anti-deaminated Gliadin peptide >143 (N<7).
- Ac IgG anti-deaminated Gliadin peptide >303 (N<7).

→ Serological profile in favour of celiac disease +++.

- Receives:
  - Gluten-free diet
  - Multivitamins
  - Peptisorb since 8.8.17.

Dietary therapy

Protein losing enteropathy

- Low fat
- High protein
- MCT

Nutrison Peptisorb

- Semi elemental food for special medical purposes: Diseases related to malnutrition in patients with malabsorption and/or maldigestion. (1 kcal/ml).
- Nutritionally complete:
- Short chain peptides and amino acids.
- Low fat; MCT 47%.
- Modified starch.
- Indication: Adults and children > 6 years.
- Can be used with caution in children 1 - 6 years.
- → → → Nutrini Peptisorb +++

Celiac disease

- Gluten-sensitive enteropathy / nontropical sprue
- Immune-mediated inflammation of the small intestine
- Sensitivity to dietary gluten and related proteins in genetically predisposed individuals (DQ2/DQ8)
- Different from food allergies, (IgE or IgG).
- Common disorder: 0.5 to 1% of general population in most countries

Gluten

- Wheat (Blé) : Prolamins

  Protein fraction of Wheat Flour :

  High content of AA: Proline & Glutamine

  - Alcohol soluble fraction: Gliadin

  - Alcohol insoluble fraction: Glutenin
- Rye (Seigle) : Secalins
- Barley (Orge) : Hordeins
- *Oats (Avoine): Avenins*

Classical symptoms of celiac disease

Symptoms of malabsorption:

- diarrhea,
- steatorrhea,
- weight loss.

Other gastrointestinal symptoms:

- abdominal pain,
- flatulence,
- abdominal distension,
- paradoxical constipation

Non-gastrointestinal manifestations

- Failure to thrive
- Persistent diarrhea
- Chronic constipation
- Recurrent abdominal pain
- Vomiting
- Dental enamel hypoplasia of permanent teeth (symmetric distribution)
- Idiopathic short stature
- Significant pubertal delay

Non-gastrointestinal manifestations (2)

- Iron deficiency anemia not responsive to supplementation
- Dermatitis herpetiformis-like rash
- Recurrent aphthous stomatitis
- Fracture not explained by the level of trauma
- Arthritis or arthralgias (even if there is a presumed underlying rheumatic disease)
- Abnormal liver biochemical tests
- Chronic fatigue

High-risk groups

- First-degree relatives of patients with celiac disease
- Autoimmune thyroiditis
- Type 1 diabetes
- Down syndrome
- Selective immunoglobulin A (IgA) deficiency
- Turner syndrome
- Williams syndrome
- Juvenile chronic arthritis

Attention

Un train peut en cacher un autre!!!


ATTENTION !!! 1 TRAIN
PEUT EN CACHER 1 AUTRE

Thank you

Case No. 2:
Lucas, 6 yrs, male

- Fever since a few days,
- Treated by parents with PCM and Diclofenac
- Pallor+++

- Hb 4.9. WBC 10 600 (N 7.37, Lc 1.7),
- Plq 135 000.
- MCH 28 MCHC 32.
- Nucleated RBC++, polychromatic cells++,
- Target cells +, macrocytes++.

Case No. 2:
Lucas, 6 yrs, male

- Acute anemia: pallor+++
- Jaundice since 3 days
- Splenomegaly ++
- Poor general condition
- Vital parameters +/-

- History: Blood transfusion at the age of 8 m.

Action?

- Private clinic
- 9.00 p.m.
- Acute severe anemia
- Vital parameters +/-
- Blood tests unavailable

Blood tests

- Cross-match
- Anemia confirmed
- Hyperbilirubinemia predominantly unconjugated
- Coombs Test negative
- Blood Smear: Malaria negative
- Hb Electrophoresis: Normal
- G6PD deficiency confirmed

G6PD deficiency

- The most common enzymatic disorder of RBCs (400 million people worldwide)
- G6PD generates NADPH and protects RBCs from oxidative injury.
- X-linked disorder
  - Males: Hemizygous
  - Females: Heterozygous

Geographical distribution

- Kurdish Jews: 60 - 70 %
- Sardinia: 4 - 35 %
- Nigeria: 22 %
- Thailand: 17 %
- African Americans: 11 - 12 %
- Brazilian Blacks: 8 %
- Greeks: 6 %
- South China: approx 6 %
- India: 3 %
- Japan and Korea: 0 to 1 %
- Strong correlation with once-malaria endemic regions

Medicines and other substances thought to be unsafe (1)

Antimalariques

- Primaquine, Pamaquine.
- Pentaquine, Quinine, Mépacrine, Plasmoquine, Chloroquine, Quinacrine.

Antipyrétiques et analgésiques

- Acétanilide, Phénacétine.
- Antipyrine, Acide acétylsalicylique, Aminophénazone (Amidopyrine), Phénazone, Phénicarbazide.

Medicines and other substances thought to be unsafe (2)

Tous les sulfamides

- Sulfalinamide, Sulfapyridine, Sulfamérazine, Sulfacétamide,
- Sulfathiazole, Sulfisoxazole et dérivés, Sulfaméthoxypyridazine.

Tous les sulfones

Medicines and other substances thought to be unsafe (3)

Tous les nitrofuranes

- Furadoïne®, Furoxane®. Acide nalidixique.

Médications diverses

- Ac para-aminosalicylique (PAS),Phénylhydrazine, Acétylphénylhydrazine, Bleu de méthylène, Dérivés de Naphtalène, Ac ascorbique, Probénicide, Quinidine, Trinitrotoluiène, Chloramphénicol, Vit K hydrosoluble, Néoarsphénamine (Collunovar®), Isoniazide. Dimercaptol.

Medicines and other substances thought to be unsafe (4)

Produits végétaux

- Fèves, certains haricots, pois verts, pois chiches, artichauts, asperges, figues de Barbarie, etc.

<u>Autres situations déclenchantes</u>

- **Infections**
- Hépatite virale, Mononucléose infectieuse, Fièvre thyphoïde.

Autres facteurs déclenchants

- Acidose diabétique

Prevention of future episodes of hemolysis

- Identification of G6PD deficiency
- Patient education regarding safe and unsafe medications and foods.

Thank you