Favism

See also separate Glucose-6-phosphate Dehydrogenase Deficiency article.

Favism describes the susceptibility to, and clinical presentation of, acute haemolytic crises as a consequence of eating broad beans in a subgroup of patients with glucose-6-phosphate dehydrogenase (G6PD) deficiency.

It can be potentially life-threatening. Broad beans are derived from the plant Vicia fava - hence, the condition's name. Susceptible patients may also experience the syndrome when exposed to the plant's pollen.

G6PD is crucial in maintaining red cell homeostasis and its deficiency leads to increased susceptibility to haemolysis induced by drugs, infections and substances in food. There is a huge number of polymorphisms of the gene with variable effects on the activity of the enzyme and a wide range of phenotypic susceptibility to haemolysis.

Genetics

The gene for G6PD is located on the X chromosome. It is therefore an X-linked inherited disease that primarily affects men. It can have clinical effects in homozygous women and a proportion of female heterozygous carriers.

It is thought that the susceptibility to favism is determined by a combination of the particular G6PD polymorphism and other factors, such as genetics and also metabolism of the active ingredients in the beans, which causes oxidative damage in red blood cells. An infectious agent such as a virus may also play a role in the development of favism.[1]

Epidemiology

- G6PD deficiency is the most common enzymopathy of man of clinical significance. It affects more than 400 million people worldwide.[2]
- Patients with favism are always G6PD-deficient but not all G6PD-deficient individuals develop haemolysis when they ingest fava beans.
- The vast majority of cases of favism occur in individuals with severely deficient variants of G6PD.
- Favism is more common in males than in females.[3]

Presentation

- There may be a past history of episodes of neonatal or childhood jaundice.
- A dietary history may reveal recent ingestion of broad beans.
- Check for recent medication changes or history consistent with infection.

Favism leads to acute, massive intravascular haemolysis. Its main clinical features are:

- Acute back and/or abdominal pain.
- Acute pallor due to anaemia.
- Haemoglobinuria causing the passage of dark or orangey-yellow urine.
- Jaundice.
- Patients with G6PD deficiency are prone to gallstones and splenomegaly due to recurrent, often subclinical, episodes of haemolysis.
Differential diagnosis

- Acute haemolysis caused by an alternative precipitant in those with G6PD deficiency - eg, drugs, particularly antimalarials or infection.
- Sickle cell anaemia and crisis.
- Exacerbation of other haemolytic anaemias - eg, hereditary spherocytosis, autoimmune haemolytic anaemia.
- Disseminated intravascular coagulation.
- Systemic lupus erythematosus.

Investigations

- Dipstick urine testing to reveal evidence of haemoglobinuria.
- FBC will show acute haemolytic anaemia picture with low haemoglobin.
- Reticulocyte count may be elevated (although often normal in the early acute phase).
- Raised indirect bilirubin (unconjugated) indicating haemolysis.
- LFTs are usually normal.
- Serum lactate dehydrogenase may be elevated, indicating haemolysis.
- Serum haptoglobins may be low, indicating haemolysis.
- Abdominal ultrasound may be used to detect gallstones and/or splenomegaly.
- Coombs' test is negative.
- G6PD activity assay in undiagnosed cases - may be normal if there is significant reticulocytosis, as reticulocytes are rich in the enzyme; assay may need to be repeated in the convalescent phase.

Associated diseases

- G6PD deficiency.
- Gallstones due to chronic haemolysis.
- Splenomegaly due to chronic haemolysis.

Management

- Avoid further ingestion of broad beans.
- Folic acid supplementation.
- Iron supplementation if there is ongoing acute severe intravascular haemolysis.
- Oxygen therapy.
- Bed rest and transfer to a high care/intensive care setting.
- Intravenous fluids to reduce the chance of acute oliguric renal impairment.
- Blood transfusion or exchange transfusion is sometimes needed to treat severe anaemia.

Complications

- Death due to acute severe haemolytic anaemia (relatively rare).
- Ophthalmological damage due to intraocular intravascular haemolysis.
- Acute kidney injury.
- Susceptibility to infection.

Prognosis

This is variable depending on the degree of susceptibility to favism, quantity of beans ingested and access to acute medical care. Most cases do well with supportive care but there is significant morbidity and some mortality associated with the disease.
Prevention

- Avoidance of ingestion of broad beans in patients known to have G6PD deficiency, or who have suffered previous episodes of favism.
- However, one study found that the majority of patients who ate fava beans after an attack had no symptoms.\(^1\)
- Genetic counselling and screening may be useful where there is a family history of G6PD deficiency, to allow diagnosis before exposure to haemolytic precipitants.
- Population screening and health education programmes in areas of high prevalence of G6PD deficiency have been shown to reduce the incidence of favism in the at-risk population.\(^4\)
- See separate article on G6PD deficiency for a list of medications to be avoided in G6PD-deficient patients.

Further reading & references


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