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Hepatomegaly and multiple liver lesions

Ho-Choong Chang, Ba Nguyen, Fintan Regan

A 33-year-old man was referred for possible liver transplantation. The patient was initially diagnosed at birth when he presented with an enlarged liver and episodes of hypoglycaemia. A liver biopsy at the time showed pale hepatic cells by virtue of cytoplasmic granularity and periportal nuclear ballooning (figure 1). He was treated initially with dietary modifications but subsequently required night time dextrose and corn starch. Failed medical therapy prompted referral for liver transplant evaluation. Physical examination showed massive hepatomegaly. Liver function tests were abnormal with a significantly raised alkaline phosphatase and transaminase. Sonography showed hepatomegaly with multiple focal lesions unchanged in size since ultrasound 3 years earlier. Computed tomography (CT) showed multiple well-defined low-attenuation lesions throughout the liver. The largest of these measured $8 \times 8 \times 4$ cm and contained foci of coarse calcification (figure 2).

Figure 1 Liver biopsy showing pale hepatic cells

Figure 2 Axial CT scan showed multiple well-defined low-attenuation lesions throughout the liver

Imaging Department,
Johns Hopkins
Bayview Medical
Centre, Johns Hopkins
University Medical
School, Baltimore, MD
21224-2780, USA
H-C Chang
B Nguyen
F Regan

Correspondence to Dr Regan

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Questions

- 1 What is the probable diagnosis, and what enzyme deficiency and type of inheritance characterise this disease?
- 2 What substance accumulates in the liver and kidney?
- 3 What liver complications are associated with the disease?

Answers

QUESTION 1

The probable diagnosis is type 1a glycogen storage disease (Von Gierke's disease). Von Gierke's disease is rare with an incidence of approximately 1 in 100 000. It is characterised by an autosomal recessive inheritance of glucose-6-phosphatase deficiency.

QUESTION 2

Sufferers from Von Gierke's disease are unable to hydrolyse glucose from glucose-6-phosphate produced from either stored glycogen or gluconeogenesis, leading to an accumulation of glycogen in the liver, kidneys, and bowel.

QUESTION 3

Hepatomegaly is common and the incidence of both adenomas and hepatocellular carcinoma is increased. Although histology of the lesions was not obtained, their stable appearances over three years made exclusion of hepatocellular carcinoma possible.

Discussion

Usually, hepatic adenomas are a spontaneous occurrence or associated with oestrogen intake. Most adenomas are single, but when associated with underlying metabolic disorders may be multiple. Hepatocellular adenomas and hepatocellular carcinoma are both complications of Von Gierke's disease¹ and are thought to stem from persistent hormonal stimulation due to chronic hypoglycaemia.² CT imaging is essential to diagnose these complications, to guide

Learning point

Although rare, liver adenomas do occur in Von Gierke's disease and the association should be recognised, as their presence may be an indication for liver transplantation

biopsy of the lesions if malignant degeneration is suspected, and to provide liver volume estimation if transplant is considered. Typical abdominal CT appearances of Von Gierke's disease include hepatosplenomegaly and nephromegaly due to glycogen deposition. Liver adenomas associated with the disease are usually low in attenuation but may be spuriously high in density if there is underlying fatty infiltration. Areas of necrosis, haemorrhage, or calcification are not uncommon. Rapid enlargement of the adenomas should suggest malignant degeneration or bleeding. Orthotopic liver transplantation has been performed to correct the metabolic defect in those unsuccessfully managed by conventional means,³ but the presence of multiple large adenomas with their risk of haemorrhage is, as in our patient, also an indication for transplantation.

Final diagnosis

Hepatocellular adenomas in association with Von Gierke's disease.

Keywords: hepatocellular adenomas; Von Gierke's disease

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Medical Anniversary

A J CRONIN, 19 July 1896

Archibald Joseph Cronin (1896–1981) was born in Cardross, Cumbartonshire, and qualified in Glasgow in 1919. He practised medicine in the Rhondda mining valleys and later converted this experience into vivid novels, biography, radio and television series. — *DG James*