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CASE REPORT

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Fatal hepatic haemorrhage in a child—peliosis hepatis versus maltreatment

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Abstract A 2.5-year-old boy with known myotubular myopathy (Spiro-Shy-Gonatas syndrome) and gonadorelin intake 9 months ante-mortem was found dead in his bed at home. At autopsy a ruptured subcapsular haematoma of the liver with resulting haemoperitoneum (600 ml) was found. Both lobes of the liver showed numerous circular blood foci <1 mm–2 cm in diameter. Signs of mechanical trauma such as bruising of the abdominal wall were absent. Histologically, the blood cysts were commonly connected to the sinusoids but did not have an endothelial lining and the reticular fibres showed ruptures. These pathomorphological findings are characteristic for peliosis hepatis and the cause of death was therefore determined to be exsanguination due to hepatic haemorrhage from peliosis hepatis instead of from mechanical trauma. To our knowledge this is the youngest casualty from peliosis reported so far.

Keywords Peliosis hepatis · Myotubular myopathy · Sudden death · Infants

Introduction

Sudden death of infants or young children may be due to natural diseases [1] or unnatural causes such as accidents [4] or maltreatment [7, 8]. We report a case where this important differential diagnosis was unclear after autopsy because the correct diagnosis was mainly based on histological examinations.

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Case report

A 2.5-year-old boy suffering from congenital myotubular myopathy (Spiro-Shy-Gonatas syndrome) was found dead in bed by his mother. Resuscitation was unsuccessful. He had spent his first 8 months in hospital and he had still required artificial respiration at night. Gonadorelin (Kryptocur, 1.2 mg/day) had been administered 9 months ante-mortem for a period of 4 weeks because of maldescensus testis.

An autopsy was ordered because the suspicion of external suffocation emerged. The body was in a well-cared for condition but severe generalized muscle atrophy was obvious. The skin and mucosae were pale and signs of mechanical trauma were absent. In particular, the abdominal skin and wall as well as the neck and face were free of bruising, excoriations or haematomas. There was 600 ml of blood in the abdominal cavity originating from the liver, which was clearly enlarged (495 g) and showed a thick subcapsular haematoma almost enclosing the whole of the right lobe. The capsule showed a rupture 8 cm in length at the upper anterior surface of the right lobe. Signs of anaemia were intense and the cause of death was exsanguination but it remained unclear after autopsy if the bleeding from the liver was caused by vital trauma, resuscitation or natural disease.

After fixation of the liver in toto the cut surfaces showed a ruptured subcapsular haematoma and the liver parenchyma was light-coloured and vulnerable. It showed a map-like pattern and both lobes of the liver were randomly interspersed by numerous circular bleedings <1 mm–2 cm in diameter (Fig. 1).

Histologically, myotubular myopathy with central myonuclei was verified. In the liver, sinusoidal ectasia and numerous tiny blood-filled spaces were diffusely distributed in both lobes (Figs. 2 and 3). The blood foci were commonly connected to the sinusoids but did not have an endothelial lining (Figs. 2 and 3). There was sometimes a topographic relation to the periportal fields, which showed a slight fibrosis (Fig. 3) and clearly dilated veins (Fig. 2). Staining for iron was negative and the

Fig. 1 Cut surface of the liver after fixation. Thick subcapsular haematoma of the right lobe and light coloured liver parenchyma which could be easily torn. Numerous small and sometimes spherical blood spaces in the liver parenchyma, some of which appear to have flowed together in the right lobe

Gomori staining for reticular fibres showed structural weaknesses including fragmentation.

The cause of death, therefore, was exsanguination due to hepatic haemorrhage from peliosis hepatis.

Discussion

The differential diagnosis of trauma versus peliosis is based on the microscopical findings in peliosis hepatis: the blood-filled spaces vary in diameter from less than 1 mm to several centimeters and are randomly distributed in the liver parenchyma, commonly affecting both lobes [10, 13, 17]. Typical characteristics further include endothelium-lined dilated sinusoids ("phlebectatic" form) or blood spaces enclosed by thinned hepatic cell cords ("parenchymal" form), widened spaces of Disse, ruptures of intrasinusoidal reticular fibres but relatively normal hepatic parenchymal cells [3, 10, 13, 16, 17].

The findings in our case clearly fulfilled these diagnostic criteria for peliosis hepatis, which was first recognized by Wagner in 1861 and named by Schoenlank [12]. It is a rare disorder initially reported in patients with wasting diseases such as tuberculosis [18] or malignant tumours [16]. Later, an association to anabolic or contraceptive steroid treatment [10, 13] was frequently reported and peliosos hepatis-like lesions have been produced experimentally in animals by administration of oxymetholone [10] and prednisolon [15]. Recent reports also suggest a possible association to rare genetic disorders such as myotubular myopathy [5, 14]. In addition, a causal connection between administration of the steroid gonadorelin (Kryptocur) and development of peliosis appears possible in the case reported.

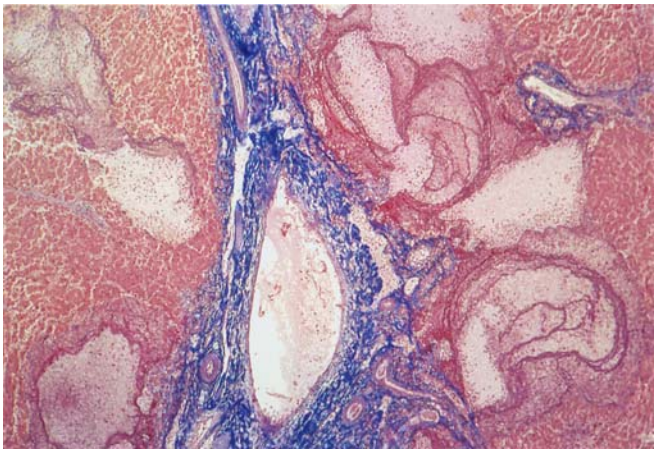

Fig. 2 Blood-filled cavities originating from dilated and ectatic sinusoids which do not have endothelium lining. Instead, the cavities are lined by thinned hepatic cell cords ("parenchymal" form). The central vein is clearly dilated. Azan, original magnification ×64

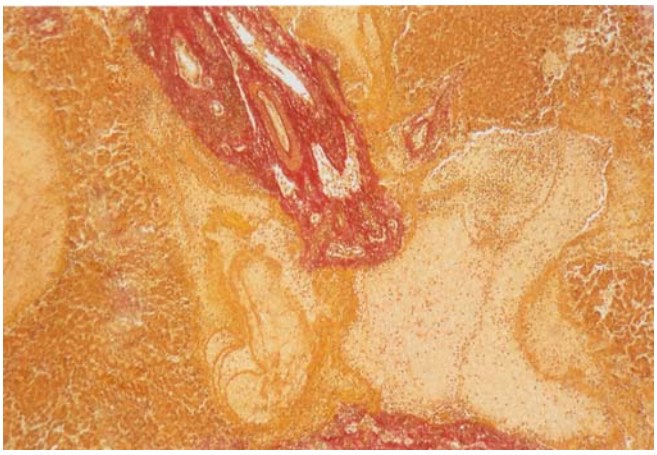

Fig. 3 Several blood spaces in close relation to the periportal fields which show a mild fibrosis. The hepatocytes appear to be normal. Sirius red, original magnification ×40

Clinically, peliosis hepatis frequently remains silent or the symptoms are uncharacteristic. Hepatomegaly or elevation of transaminase levels may occur [10, 13] but the roentgenographic diagnosis is difficult [9, 11, 14] and severe complications such as acute hepatic failure or intraperitoneal haemorrhage occur infrequently so that the diagnosis is mostly an incidental finding at autopsy [13, 14]. In children, hepatic haemorrhaging from peliosis is an extremely rare event [2, 6, 14] and to our knowledge this is the youngest casualty from peliosis hepatis reported so far.

This case report once more demonstrates that all infants and children should be autopsied even if there is no indication of relevant medicolegal findings in the beginning of a death investigation. Mechanical trauma does not necessarily produce external signs because organ injuries and fractures can be hidden under the inconspicious skin in infants and children [7, 8]. On the other hand, autopsy results have to be evaluated carefully and should be supplemented by additional investigations such as toxicology and histology. Abdominal haemorrhages in a child, for example, are highly suspicious of trauma but this case illustrates that spontaneous bleeding due to natural disease may also occur.

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