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Hepatic Vein occlusion Disease: Report of a Case in the North Central State of Nigeria

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Summary

Edington, G. M. and Haggie, M. H. K. (**1977**). *Nigerian Journal of Paediatrics*, **4** (**1**), 6. **Hepatic Vein occlusion Disease: Report of a Case in the North Central State of Nigeria.** The clinical features and laboratory findings are described in a child suffering from a sudden onset of hepatomegaly and ascites. The histopathology of the liver biopsy considered by us as unusual, is described in detail. Attention is drawn to the fact that superficially this type of hepatic lesion resembles that due to chronic venous congestion. A possible causal relationship with plant alkaloids is suggested.

A CENTRAL form of hepatic cirrhosis in children has long been known to occur in Jamaica (Mcfarlane and Branday, 1945; Hill *et al.*, 1953). The last named authors discussed the pathogenesis of the hepatic fibrosis under the title of "serous hepatosis". Brass, Jelliffe and Stuart (1954) named the same condition 'veno-occlusive disease', and considered it to be due to obliterative lesions of the medium-sized and smaller ramifications of the hepatic veins with non-portal cirrhosis as the ultimate result. The lesions in the hepatic veins were considered to be due to the ingestion of herbal infusions containing alkaloids from the plant *Crotalaria fulva*. The authors also claimed that Species of *Senecio* could produce a similar condition. Allen, Carstens and Katagiri (1969) injected monkeys with monocrotaline and noted fragmentation of the endothelium of the small and medium-size hepatic veins with oedema of the vessel wall and ultimately obliteration of the vascular lumina. Brooks *et al*, (1970) in an ultrastructural study of the liver in six Jamaican children with acute veno-occlusive disease described endothelial damage in the sinusoids, subterminal and terminal hepatic veins with extravasation of erythrocytes into the space of Disse.

Veno-occlusive disease has also been described from other parts of the world. In South Africa, Selzer and Parker (1951) have described 12 cases presenting as Chiari's syndrome, the authors attributed the condition to *Senecio* poisoning. Stein (1957) also reported four cases in children aged two years and under. In Iraq, Al-Hassany and Mohamed (1970) noted the condition in 9 children under 12 years of age. In all these patients the centrilobular and sublobular hepatic veins were affected, and plant alkaloids were considered as possible aetiological substances. By contrast, Safouh and Shehata (1965) from

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Egypt, and Grases and Beker (1972) from Venezuela have described cases in which the primary involvement appeared to be thrombosis of the main and medium sized branches of the hepatic vein with the centrilobular veins not uniformly affected. These authors also suggested that plant alkaloids were involved in the aetiology of the condition. This entity which Safouh and Shehata (1965) called 'hepatic vein occlusion disease of children', should indeed, be included among the causes of the Budd-Chiari syndrome. Gibson (1960) has drawn attention to the difference between Chiari's disease in which primary occlusion of the ostia of the hepatic veins occurs, and the Budd-Chiari syndrome in which occlusion of the hepatic veins is the presenting lesion. There are many causes of the Budd-Chiari syndrome, but in temperate climates, young adults are usually the victims. An interesting feature of Gibson's cases of Chiari's disease was the presence of numerous small fibrous tags on the surface of the liver. In both Chiari's disease and the Budd-Chiari syndrome the pathology of the liver is said to resemble that of cardiac cirrhosis.

Veno-occlusive disease has not, to our knowledge, been diagnosed in Nigeria although it is known that plant alkaloids are widely used in folk medicine. Accordingly the following case is considered worthy of publication as the histopathological findings in the liver were unusual and have not previously been seen by one of us (G.M.E.) in the examination of many surgical biopsy and necropsy specimens in 30 years of practice in West Africa.

Case Report

A Hausa girl, aged 4 years, was admitted to the Ahmadu Bello University Teaching Hospital, Zaria, with a history of 4 weeks diarrhoea and three weeks abdominal swelling. She had been given some local medicine at the onset of the illness but no further details could be obtained. The past medical history was non-contributory.

On examination, she was small and thin for her age, but in good general condition and had no jaundice or anaemia. There was massive ascites. The liver was enlarged to 10 cms below the right costal margin, and was hard but smooth. The spleen was also enlarged to 6 cms below the costal margin; it was firm. The significance of the splenomegaly is doubtful since this is a usual feature in children gaining immunity to malaria.

Peritoneocentesis yielded clear ascitic fluid which contained no cells, but had a protein content of 2.8 gm per cent. Other laboratory findings were: haemoglobin, 13.5 gm per cent; white blood cells, 5,100 per cu. mm; Hb genotype, AA; alkaline phosphatase, 5.4 K.A units. Serum bilirubin was less than 14 mmol./litre; thymol turbidity was 2 units, and flocculation tests were negative. Rectal snips showed no schistosome ova.

Laparotomy confirmed the enlarged but smooth liver which showed no other abnormality apart from a raised portal venous pressure. A wedge biopsy of the liver was taken for histopathological studies. The ascites recurred postoperatively. Unfortunately the child was soon after removed from hospital by the parents against medical advice and has therefore been lost to follow up.

Histopathology of the Liver

The biopsy was fixed in 10 per cent neutral buffered formol-saline and routinely processed. The striking feature on haematoxylin and eosin section was the intense centrilobular congestion which superficially resembled the appearances of a liver with chronic venous congestion. However, the centrilobular and sublobular veins were dilated and empty, and compressed sinusoids containing few red blood cells were present. Towards the periphery of the lobules the sinusoids were widely dilated and trabeculae, packed with red cells, could be easily distinguished (Figs 1 and 2). The centrilobular and sublobular veins showed no endothelial necrosis, intimal oedema, or proliferation. The walls appeared

Fig. 1 *Dense hepatic centrilobular congestion with narrow empty sinusoids. Dilated, relatively avascular sinusoids in the periportal areas. The trabeculae are grossly congested (H & E x 60).*

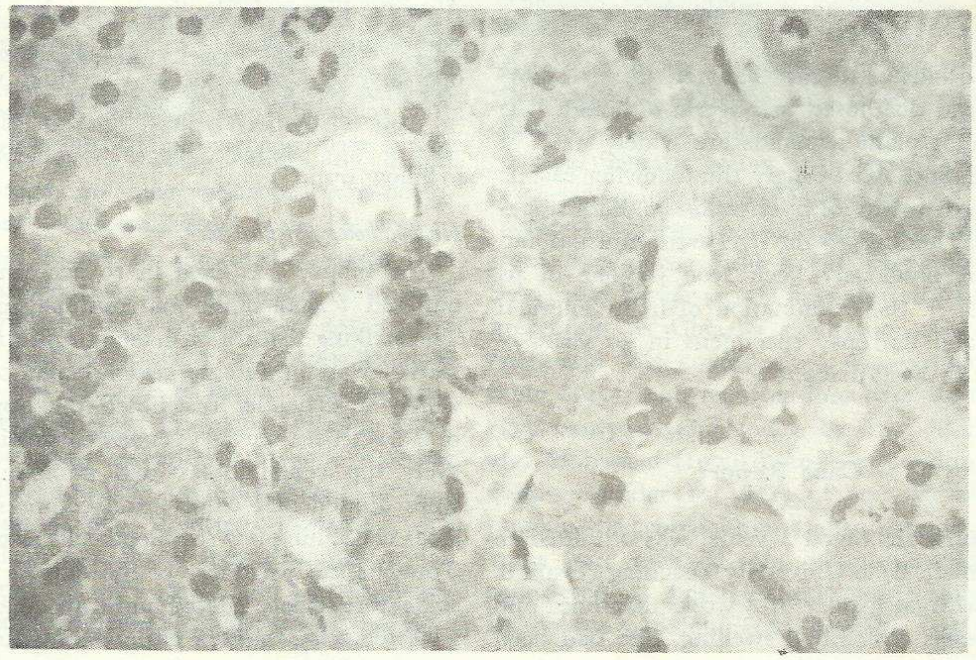

Fig. 2 *Dilated avascular hepatic sinusoids. The trabeculae are grossly congested, with loss of hepatocytes (H & E x 500).*

thickened and fibrous as revealed by reticulin and Van Giesen's stains. The significance of this was difficult to assess in the subcapsular region of the liver. A reticulin stain confirmed the collapse of the central sinusoids and package of the trabeculae with red blood cells with corresponding loss of hepatocytes. There was scattered malaria pigment in a few Kupffer cells and considerable amounts in the portal tracts which is typical of a child gaining immunity to holoendemic *P. falciparum* malaria (Edington and Gilles, 1969). No haemosiderin pigment was present. The findings were considered consistent with the diagnosis of a hepatic venous outflow block.

Comments

The clinical features in this patient were consistent with a sudden thrombosis of the main radicles of the hepatic vein. The diarrhoea could have been caused by intestinal congestion due to the raised portal venous pressure. The histopathological findings in the liver do not resemble those of chronic venous congestion, the most severe example of which in Nigeria is associated with right-sided endomyocardial fibrosis. Neither do they resemble the hepatic appearances in other heart diseases, nor the classical hepatic findings in veno-occlusive disease in which centrilobular and sublobular veins are affected. The histopathology of the liver in the present case is however similar to that described by Leopold, Parry and Storring (1970) in Welsh adults suffering from hepatic vein thrombosis in which dissection of blood into the liver trabeculae with the setting up of an extra sinusoidal circulation was evident. This condition has been further discussed by the British Medical Journal (Editorial, 1971).

Other causes of hepatic venous outflow block in the present case cannot be completely excluded. In view of the sudden onset of the illness, it is justified to consider that the child suffered from the type of liver disease thought to be due to the ingestion of plant alkaloids as described by Safouh and Shehata (1965), and by Grases and Beker, (1972). It should be noted however, that in neither of the above publications was emphasis laid on the presence of congestion of the hepatic trabeculae. It is therefore considered necessary that attention should be drawn to this type of acute liver lesion, which superficially resembles, and could be mistaken, for chronic venous congestion.

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