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EDITORIAL

A NEW DYNAMIC APPROACH TO THE DIAGNOSIS OF SYMMERS' FIBROSIS IN SCHISTOSOMIASIS BY ULTRASOUND

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Pathological aspects

The English pathologist Symmers, who was working at the Kass-El-Ainy Hospital in Cairo, described for the first time in 1904 a new form of cirrhosis of the liver caused by eggs of schistosoma[32]. From post-mortem examinations of diseased livers Symmers described lesions which he likened to "white clay-pipe stems" at various angles throughout the liver. What we nowadays call liver fibrosis[26], Symmers called cirrhosis and made the further mistake, understandable at the time, of describing what were surely **S. mansoni** eggs presenting lateral spicules in the hepatic tissue as "the ova of Bilharzia haematobia". Spindle cells arranged concentrically around the eggs were ringed with fibrous tissue that formed "periportal cirrhosis".

Subsequently pathologists in various countries, mainly Egypt[20, 29], Puerto Rico[25] and Brazil[3, 4, 7, 8, 11, 14] have used the terminology coarse periportal fibrosis[7, 20], axial fibrosis[12], pipe stem fibrosis[3], or the best-known term, Symmers' fibrosis, in dealing with the hepatopathy of advanced Bilharziasis.

Brazilian pathologists from the city of Recife (COUTINHO[14], COELHO[12], COELHO et al.[11] and MAGALHÃES FILHO[28]), from Belo Horizonte (BOGLIOLO[7, 8]), and from Salvador (ANDRADE[3], ANDRADE & CHEEVER[4], ANDRADE & BINA[5]) have all stressed the significance of Symmers' fibrosis in both liver biopsy and post-mortem examinations. It is key to the diagnosis of advanced schistosomiasis liver disease whose morphology characterizes the hepatosplenic form of the illness. It is beyond the scope of the present work to go into the detailed macroscopic and microscopic description of Symmers' fibrosis, let alone its much-disputed pathogenesis[6, 19], which are well discussed in the literature cited above.

Post-mortem examinations particularly would indicate that periportal fibrosis, or Symmers' fibrosis, is almost always present in advanced forms of **S. mansoni**. Such were the findings of ANDRADE & BINA[5], in their review of 232 post-mortems in the Hospital das Clínicas da Bahia in the city of Salvador, Brazil. We take the liberty to show, in Fig. 1, an original illustration by Symmers himself. Comparison with Fig. 2, kindly supplied from his files by Prof. Zilton Andrade, makes the point.

Symmers' fibrosis is also frequently mentioned in wedge biopsy reports on hepatosplenic patients who have undergone surgery, as the 85.5% in Recife[24], or laparoscopy examination[30]. This diagnosis is rare, however, in percutaneous needle biopsies, where the specimens are small. But this latter histology is useful in schistosomiasis in the analysis of other aspects, including post-treatment progress case studies[15].

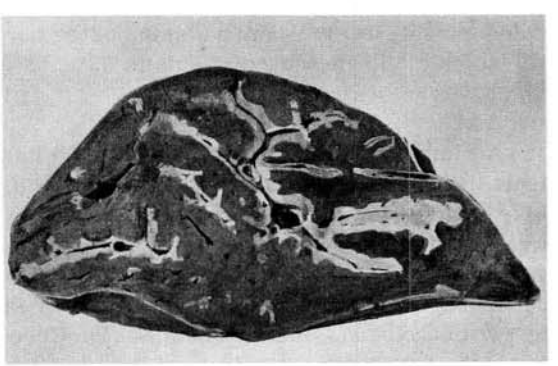

Fig. 1 — "Longitudinal section of portals canals, in which there is great increase in the amount of fibrous tissue around the vessels and bile ducts". (Symmers, 1904).

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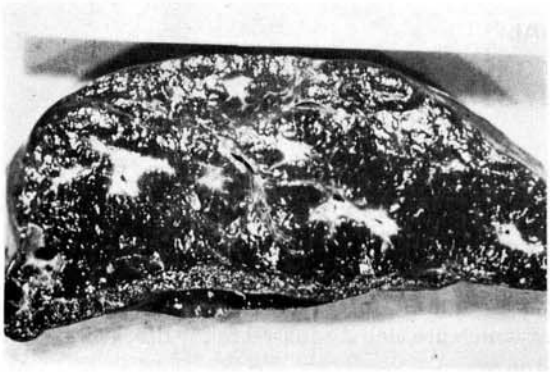

Fig. 2 — Symmers pipe-stem fibrosis from the collection of Prof. Zilton Andrade.

Ultrasound aspects

Diagnosis of liver diseases has benefited during the seventies and eighties from ultrasound technology, particularly in malign[15] and benign[17] focal lesions, and in the study of cholestasis. Less so in the case of diffuse liver diseases such as cirrhosis[27]. Recently, however, state-of-the-art ultrasound has contributed greatly to the study of liver fibrosis and portal hypertension[16], particularly from **S. mansoni**[1, 9, 10, 18, 23, 31].

Research by CERRI[9] and CERRI et al.[10] into 103 patients with the hepatosplenic form of the disease in the city of São Paulo, Brazil, showed periportal fibrosis in 73%, hypertrophy of the left lobe in 81%, gall bladder wall thickening in 60%, splenomegaly in 100%, and widening of the portal, splenic and mesenteric veins in 73%, 68% and 42% of cases, respectively.

An important step forward in this field has recently been made by HOMEIDA et al.[21, 22], working in the Sudan. In their first study[21], they were looking for a correlation between pre-surgery ultrasound data and the results of histology specimens from surgical biopsies as examined by two pathologists, among whom was Dr. Cheever of the US National Institute of Health. High correlation was found in 41 patients with Symmers' fibrosis and in ten patients without Symmers' fibrosis in a hospital in Khartoum between conventional pathology diagnosis and ultrasound findings.

They accordingly proposed a four-level classification of the extent of periportal fibrosis as revealed by ultrasound. No patient, however has been found in this hospital study to show the first postulated grade of the fibrosis. They came to the conclusion that ultrasound is at least as sensitive a diagnosis technique in Symmers' fibrosis as is wedge biopsy. And, as ultrasound is a non-invasive technique, it should be the method of choice for this diagnosis.

In their second study[22], HOMEIDA and others carried out ultrasound diagnosis, without liver biopsy, in field studies at two villages in Gezira, Sudan. Groups of 353 and 247 people were selected at random from the two total populations of 1,210 and 1,000. Symmers' fibrosis was diagnosed by ultrasound in 18% and 13% respectively of the two groups, in whom infection with **S. mansoni** was found in 71% and 37%. These cases were classified into three grades of fibrosis. In contrast to the first study, in which no one was found to show grade 1, this second field study showed grade 1 fibrosis in 60% and 65% of the two groups of 65 and 32 people with Symmers' fibrosis. This grade 1 subgroup was judged by the authors as showing the initial phase of the disease, or possibly a stabilized older condition, or else the fibrosis in the process of regression.

Grades 2 and 3, representing 34% and 32%, and 6% and 3% respectively in each village, showed more advanced stages, such as are commonly met with in hospital sample populations[21]. On the other hand, the prevalence and degree of splenomegaly in the second study, as equally the diameter of the portal and splenic veins, indicating portal hypertension, correlate well with the extent of liver fibrosis. This is not the case with hepatomegaly.

The presence of Symmers' fibrosis as diagnosed by ultrasound was according assessed by HOMEIDA et al. as two to three times the clinical prevalence of splenomegaly and of hepatosplenic schistosomiasis disease. The authors in their second field study believed then that Symmers' fibrosis was clearly shown to be much more common than had been previously thought. This is, in our view, patently so, as long as the diagnosis of Symmers' fibrosis as slight, or grade 1, is generally accepted. For it to be so would

demand fuller data from observers in other endemic areas following more objective criteria supported by the corresponding histopathology studies. HOMEIDA et al. description of grade 1 is fairly subjective, and imprecise: "minimal echogenic thickening of the walls in ≥ 2 of the portal vein radicles with little change in the diameter of the main portal vein, or diffuse thin, linear echogenicity scattered over the liver".

When the present review was almost complete, we had the opportunity to read a January' 89 paper by ABDEL-WAHAB et al.[2] working in Egypt. They describe their findings from ultrasound in the hospital, which they compare with pathology results from surgical liver biopsies in the diagnosis of schistosomiasis fibrosis, cirrhosis, and combined lesions. Although the Egyptian researchers did not classify the degree of Bilharzia liver fibrosis as had HOMEIDA et al., they stress the importance of ultrasound in diagnosing gross lesions of Symmers' pipe-stem fibrosis. They found it less certain in showing the initial lesions usually met with in asymptomatic children.

In the case of advanced lesions, we believe that ultrasound, which can show ample areas of the liver and clear signs of portal hypertension, could with advantage replace the more restricted area dealt with in a wedge liver biopsy and particulary in a needle biopsy. Or at least become a valid alternative. Moreover, ultrasound is harmless, simple and non-invasive. It can, accordingly, be repeated periodically in the same patient. A more accurate first hand experience is shortly expected from our current work under way at the Hospital das Clínicas of the Universidade Federal de Pernambuco with Dra. Ana Lúcia C. Domingues, and at the Aggeu Magalhães Research Center with Dra. Ana Regina F. C. Lima, among others. This research embraces clinical aspects, endoscopy, pathology and ultrasound (Fig. 3) in hospitalized hepatosplenic schistosomiasis patients, to hopefully be extended to include field work.

This brief review supports the view that ultrasound examinations are of real importance in current research into the morbidity of **S. mansoni**, and of significance in a better knowledge of the natural history of the disease. We have, however, reservations about the adequacy of

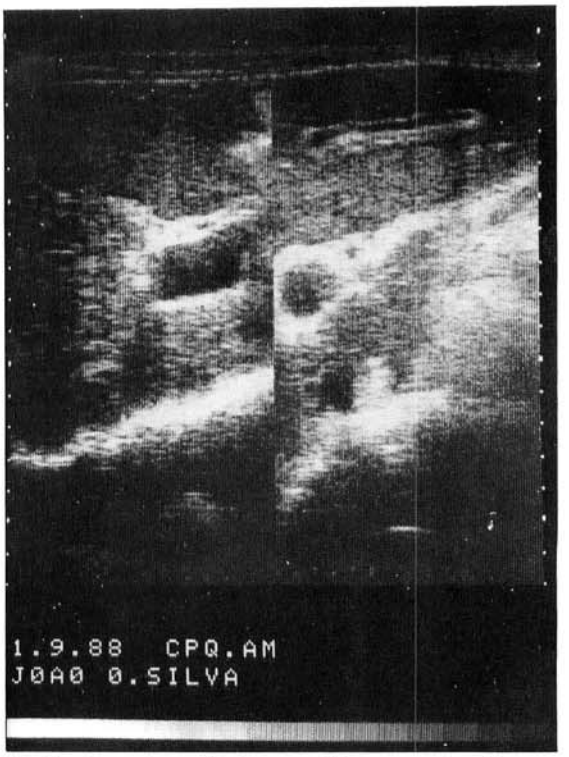

Fig. 3 — Two echographic images of the same patient showing periportal fibrosis and enlargment of the portal vein (Dra. Ana Regina Lima, Recife).

Symmers' fibrosis denomination for grade 1 as used by HOMEIDA et al. ABDEL WAHAD et al. judged this degree difficult to classify. And Symmers' own original description, confirmed by all subsequent pathologists, was of a much more intense and widespread fibrosis. A simple general denomination of portal fibrosis or, better, periportal fibrosis, classified in three different grades, might be more suitable and would be of use in both pathology and ultrasound studies, keeping the term Symmers' fibrosis for its historical value. This would be more in line with the pathologists who have well studied experimental or human schistosomiasis pathology, such as Zilton Andrade, Jean Alex Guimaud, Allen Cheever, F. Lichtenberg and Kenneth Warren, among other eminent experts.

At all events, ultrasound opens new horizons and throws fresh light on the study of liver fibrosis of schistosomiasis origin in man, in a new, dynamic, approach.

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