Portal Cavernoma Cholangiopathy

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In this issue of the journal the reader will find a bunch of articles that represent an unprecedented attempt at clarifying and simplifying our views on the bile duct anomalies that are related to portal cavernoma, the major variant of chronic extrahepatic portal vein obstruction. For this purpose, the Indian Association for the Study of the Liver organized a consensus conference that took place in Chandigarh on 12th and 13th October 2012. Indian experts convened for presenting their review of the literature data on various aspects, from nomenclature to etiology, pathogenesis, manifestations, diagnosis, treatment and outcome. After an extensive discussion of these aspects, they elaborated a consensus statement based on their unequaled experience in the management of this rare condition. Furthermore, articles corresponding to the individual expert surveys of the literature and experience have been elaborated, extensively discussed and reviewed by the editors. Much emphasis was given not to escape any necessary discussion and confrontation of different opinions coming from different viewpoints. However, attention was also given to reach a comprehensive and consistent view of the condition and the issues it raises for clinical practice. The Editors believe that the result is outstanding and hope this appreciation will be shared by the reader.

Among the most outstanding achievements has been reaching a consensus to adopt a new denomination, portal cavernoma cholangiopathy that gets rid of all the limitations of previous terms used for this entity, including portal cholangiopathy and portal biliopathy. Set alone, this new denomination makes very clear the entity so designated, namely the bile duct anomalies related to portal cavernoma. Bile duct anomalies observed in patients with cirrhosis or portal hypertension in general are thus definitely excluded from this entity which is highly relevant from a clinical practice point of view.

The care with which the pathogenesis of this entity has been discussed must also be highlighted. Several possibilities are presented, each with their arguments and with their direct

clinical implications: compression and deformation by the cavernomatous veins – which should be reversible following decompression of the portal venous system; but also ischemic fibrosis of the bile duct wall – which should be largely irreversible; and compression/encasement of the bile duct into a tumor-like cavernoma.

In relation to pathophysiology, the anatomy of the venous drainage of the extrahepatic bile ducts has been described extensively, based on dissection but also on imaging studies using endoscopic retrograde cholangiography or magnetic resonance cholangiography or endoscopic ultrasonography. This part is extremely original and the reader will not find in any other publication such a comprehensive and richly illustrated description of the complex venous drainage of the bile ducts in the normal and diseased states.

Manifestations, be they clinical or laboratory, make a clear part of what is and what is not clinically relevant. Indeed, it is surprising that such obvious anomalies of the large bile ducts have in many patients such a mild or absent clinical expression. A severe form of the disease is distinctly rare but when present, potentially devastating. The predictors for the occurrence of manifestations as explored by several studies have been presented. The big question that will require additional studies is whether this is a progressive disease or the biliary changes develop early in the course of portal vein obstruction and thereafter remain relatively fixed and stable.

Treatment for symptomatic portal cavernoma cholangiopathy is extremely challenging, as two players have to be considered, the bile ducts and the portal venous system. This is presented most clearly from several points of view, endoscopy, surgery, and liver transplantation, all of which potentially have a complementary role to play in different situations. Another remarkable achievement of this conference is to present the reader a global view where all treatment means are articulated.

The Editors are indebted to the authors for their exceptional work and compliance to the demanding rules of a collective work. They are confident that this will be to the marked benefit of all patients with portal cavernoma cholangiopathy and their caregivers, wherever they are living in the world.

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