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Case Report

Caroli's syndrome in a Shami goat

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Abstract

Caroli's syndrome is a congenital hepatic disorder that is characterized by multifocal, segmental dilatation of large intrahepatic bile ducts and congenital hepatic fibrosis. Diagnosis of Caroli's syndrome with polycystic kidney disease was made in a Shami goat kid based on the gross and histopathology of the hepatorenal lesions. Grossly, the liver was enlarged and diffusely greenish with pinpoint whitish foci. Both kidneys exhibited multiple renal cysts. Histopathologically, the liver exhibited multifocal hepatic fibrosis, bile duct ectasia and hyperplasia that were associated with diffuse cystic dilatation of medullary renal tubules of both kidneys. These hepatorenal lesions resemble Caroli's syndrome in human. To the best of our knowledge, this is the first report of Caroli's syndrome in a Shami goat.

Keywords: Caroli's syndrome; congenital hepatic fibrosis; polycystic kidney; goat

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Introduction

Caroli's syndrome is a congenital hepatic disorder characterized by multifocal, segmental dilatation of large intrahepatic bile ducts that are associated with congenital hepatic fibrosis (Desmet, 1992). In human, up to 60% of the patients, Caroli's syndrome is associated with polycystic kidney disease (Yonem and Bayraktar, 2007). The pathogenesis of this syndrome is not completely understood but ductal plate malformation at different levels of the intrahepatic biliary tree is the most acceptable theory (Kabra et al., 1995). At the molecular level, Caroli's syndrome is linked to fibrocystin which is a large protein encoded by polycystic kidney and hepatic disease 1 gene (PKHD1) (Menezes and Onuchic, 2006). In human, PKHD1 is expressed in kidneys, liver, and pancreas (Menezes and Onuchic, 2006). Caroli's syndrome has not been reported in Shami goats. This report deals with the gross and histopathological findings of the first case of Caroli's syndrome in a Shami goat kid.

Case presentation

A 5 d old dead Shami goat kid was presented to the Veterinary Health Centre (VHC) at Jordan University of Science and Technology (JUST). According to the owner, the animal was born normal. After 48 h, the animal stopped nursing, started to show abnormal gait, diarrhoea and weakness and finally was unable to stand. On the fifth day the animal died.

Gross examination

The body condition of the animal was poor. There was mild post-mortem autolysis. The liver was moderately enlarged and diffusely greenish with pinpoint whitish foci throughout the liver. Both kidneys were moderately enlarged with multiple renal cysts up to 8 mm in diameter. No other gross abnormalities were seen.

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Representative tissue sample were obtained and fixed in 10% neutral buffered formalin, processed and stained with hematoxylin-eosin and examined under microscope.

Histopathological findings

The normal architecture of the whole liver was disrupted with biliary dilatation, hyperplasia and portal fibrosis. These bile ducts were irregular and exhibited variable degrees of dilatation (bile duct ectasia), lined with one layer of columnar cells and less frequently with cuboidal cells. Most of these ducts contained amorphous eosinophilic material in their dilated lumina (Fig. 1). Protrusions of the duct wall into the lumen were present. Occasionally, in more advanced lesions, these protrusions formed bridges across the dilated bile ducts lumina separating the dilated bile ducts into more than one compartment (Fig. 1). Moderate fibrosis surrounded the bile ducts (portal fibrosis) was present throughout the liver sections. In multiple areas, the fibrosis breached the hepatic limiting plate and extended from one portal area to another or from one portal area to a centrilobular area (bridging fibrosis). These fibrotic areas contained numerous small bile ducts. Randomly, there were multiple areas of hepatic necrosis with moderate neutrophilic aggregates. No regenerative hepatic nodules were present.

Both kidneys showed multiple cystic dilatations affecting the medullary, cortico-medullary and occasionally the cortical renal tubules sparing and compressing most of the cortical tubules and the glomeruli (Fig. 2). These tubules were clear and lined with one layer of cuboidal cells. A few dilated tubules were filled with homogenous eosinophilic material (hyaline cast) or granular casts.

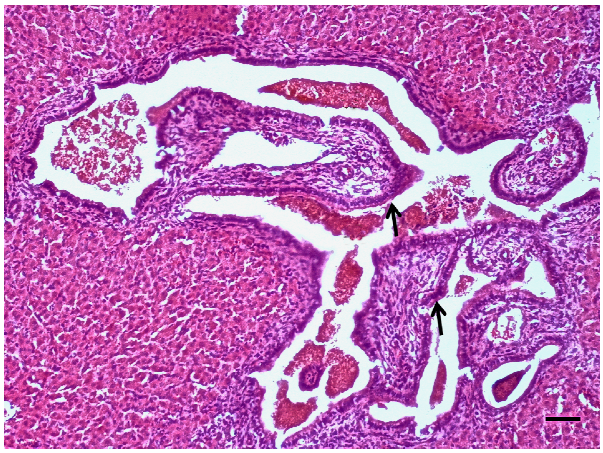

Fig. 1: Protrusions of the bile duct wall formed bridges across the dilated bile ducts lumina that were separated into compartments (arrows). The ducts contained amorphous eosinophilic material in their dilated lumina. H&E. Bar= 50µm.

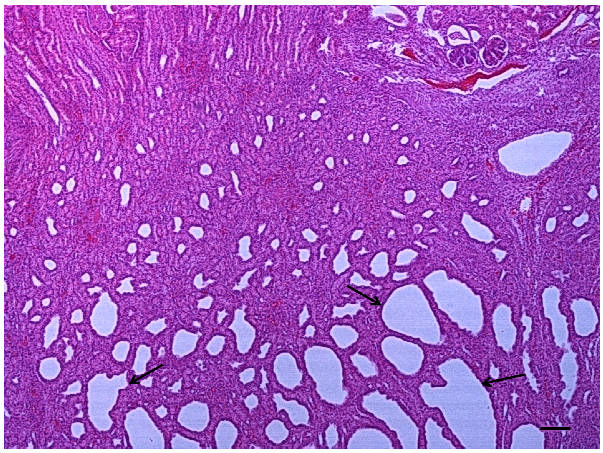

Fig. 2: The renal tubules exhibited multiple cystic dilatations (arrows). H&E. Bar= 100µm.

Discussion

The present study is the first report on Caroli's syndrome in Shami goats. Histopathological lesions in kidneys and liver were similar to those hepatorenal lesions reported in the polycystic kidney (PCK) rat model of Caroli's disease associated with congenital hepatic fibrosis in human (Sanzen et al., 2001). Histopathological findings of protrusions of the duct wall to the lumen and bridge formation of the duct wall across the lumen were suggestive of ductal plate malformation. Ductal plate malformation has been reported in dogs (Brown et al., 2010), non-human primate (Wallace et al., 2009), cats (Zandvliet et al., 2005), calf (Yoshikawa et al., 2002) and foals (Haechler et al., 2000). Hepatic portal fibrosis with frequent bile ducts ectasia and a preserved hepatic lobular architecture is known to be a feature of Caroli's disease. The renal lesions of the Shami goat kid include polycystic dilation of the renal tubules. Most of the affected tubules were at the renal medulla and cortico-medullary junction. Those lesions were similarly reported in human with such condition (Gupta et al., 2006; Kim et al., 2006).

In summary, the present case report revealed that the hepatic and kidney changes of the Shami goat kid were very similar to those lesions of Caroli's disease in human and rat model of Caroli's disease (Sanzen et al., 2001; Gupta et al., 2006; Kim et al., 2006; Wang et al., 2008). This case represents is one of the rare veterinary cases of Caroli's syndrome and the first in a Shami goat kid.

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