Adult Polycystic Liver and the Meyenburg Complexes
Biliary Ectasia

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Single case of polycystic liver in 44 years old female is reported. The lesion, microscopically, comprised of cystic channels occupying portal septa as well as hepatic lobules. Mature hepatocyte lining of the intralobular cysts may be considered a feature of adult polycystic liver. Findings suggested the origin of the cysts from the bile canaliculi, canals of Hering, interlobular ductules and the bile ducts. The Meyenburg complexes in the present case were related to the interlobular bile ductules.

Key words: Polycystic liver, Meyenburg complexes, Cysts, Ectasia, Biliary channels.

Introduction:

Meyenburg complexes as the progenitor ducts of the adult polycystic liver have been widely acknowledged, but their relationship with the functioning biliary apparatus remains hypothetical (1,2). Meyenburg's (3) view that the complexes comprised of aberrant intra-lobular ducts have been acceptable till date. Recently, however, in one polycystic liver, the possibilities of congenital bile duct ectasia could not be excluded (4). The case documented here, presented further evidences,
confirmatory of the biliary ectasia in the pathogenesis of adult polycystic liver.

Materials & Methods:

B.M.P., 44 yrs., presented with dull ache right hypochondrium and frequent vomitings and loose motions for the last 2 years. The liver was palpable, 4 fingers in the mid-clavicular line. On exploratory leproty the liver was found to be diffusely cystic. Occasional cysts were present in both the kidneys, spleen and pancreas. Few liver cysts were biopsied for examination, which revealed interesting histopathological findings. The pieces obtained in different planes were processed and several sections from each block were studied to obtain further informations about adult polycystic liver.

Observations:

On gross examination, the resected mass showed multiple cysts, the larger ones measuring 2–3 cm in diameter. The contents were transparent and serous. Microscopic examination revealed the lesion to be consisting of round, fusiform and tubularly cystic channels. These varied in size from dilatation of few microns to cysts covering several microscopic fields. The cystic channels occupied the portal septa as well as the hepatic lobules.

In the portal septa (figs. 1, 4) the bile ducts lined by columnar epithelium were observed to be branching into cuboidal lined bile ductules in the vicinity of the hepatic lobules. Some of these terminating bile ducts revealed focal fusiform or round cystic dilatations, alternating with stenotic segments. Among the bile ductules, occasional ones were found to be stenotic and lined by flattened cells immediately following their origin from the bile ducts. Majority of the portal cysts were found to be associated with these stenotic ductules and were lined by flattened cells. From 1 to 9 tributaries of each anomalous ductule revealed cystic anomalies and presented, in cross-sections, with closely placed cysts characteristics of the Meierburg complex. The complexes in the vicinity of the bile ducts comprised of columnar and cuboidal lined channels and cysts. Towards the portal-lobular junctions and in the thin portal septa, channels consisted of the inter-lobular ductules, which were lined by flattened and low cuboidal epithelium like wise their cysts.

Inside the hepatic lobules, the canals of Hering (postnatal intra-lobular ducts) corresponding to the anomalous bile ductules were found to be dilated, at places, in groups (figs. 1, 4) forming tubulo-cystic complexes and others solitarily (figs. 2, 4). These cysts were at places lined by flattened cells and in some parts by the mature hepatocytes. Occasional intra-lobular cysts occupied the deeper parts of the lobules. These cysts were lined by hepatocytes. The lobular cyst peripherally communicated with the canal of Hering (figs. 2, 4) and centrally continued into a tubularly dilated channel lined by hepatocytes which terminated as blinded loop close to the centrilobular vein (figs. 3, 4). Hepatic parenchyma around these cysts was compressed and revealed no sign of bile stasis.

Discussion:

Cystically and tubularly dilated
Fig. 1 – Tissue from the liver, showing a) fusiform cystic dilatations alternate with stenotic segments of the bile duct, b) stenotic bile ductules lined by flattened cells c) the flat cell lined portal cysts associated with the stenotic ductules. A group of cystically dilated canals of Hering d) are being seen in the parenchyma. (H & E x 50).
Fig. 2 - Tissue from the liver, showing c) portal cysts, m) Meyenburg complex comprising of branching inter-lobular ductules in a thin portal septum e) cystically dilated interlobular ductule communicating with (d) canal of Hering. The dilated canal of Hering is being seen further communicating with (f) a deep intra-lobular cyst. (H & E x 100)
Fig. 3 – Tissue from the liver, showing (f) a deep lobular cyst and (g) centrilobular tubular extension of the cyst. (H & E ×100).
Fig. 4 – A semi-diagramatic presentation of the anomalous biliary channels, showing A) cystic segments of the bile ducts, B) anomalous bile ductules, ductular tributaries (1–9), C) cystic segments of the ductules and of the ductular tributaries, cystically dilated canals of Hering and the deep intra-lobular cyst (D & F).
channels deeply penetrating the lobular parenchyma have been earlier reported and considered to be determinants of the childhood variant of polycystic liver (5). Findings were similar in the present case but differentiated on the basis of their linings. In the first case lining of columnar or cuboidal cells is a constant feature while the latter showed lining by mature hepatocytes, which, therefore, was considered to be primary evidence of adulthood polycystic liver. The features of the deep lobular cysts i.e. the hepatocytic lining, their continuity with the canals of Hering and terminations centrally close to the central lobular veins, suggested their origin from the bile canaliculi. The canaliculi in the embryonal life are tubular and, in cross sections, surrounded by multiple cells (6). Further developments lead to orientations of the hepatocytes into one or two cell thick laminae and the canaliculi are transformed into network to polygonal meshes surrounding the individual liver cells (7,8), characteristic of the adult liver. It is interesting to note that this, latter, “adult pattern” does not completely develop until a few years after the birth and under certain pathological conditions may revert back to a tubular configuration (8). In adult polycystic liver, therefore, such tubular canaliculi may be dilated from cysts of varying sizes. To best of our knowledge the canicular cysts and tubule-cystic pattern have not been earlier described in adult polycystic liver.

Morphological evidences depicting involvement of the canals of Hering in adult polycystic liver have been earlier documented by Melnick (9). In one adult polycystic liver he observed that some of the portal Meyenburg ducts presented as narrow and elongated outgrowths from the ends of the liver cell cords. Melnic proposed that in due course of time these ducts detached from the liver cords and gave rise to aberrant cysts and Meyenburg complexes in the portal septa. In the present case, terminal ends of the liver cords (post natal intra-lobular ducts), communicating with the Meyenburg (portal) ducts, were cystically dilated and formed intra-lobular solitary cysts and cyst-complexes. These cysts differed from those of Meyenburg cyst complexes in being separated from each other by hepatic parenchyma cells instead of the fibrous tissue. Their lining was additionally by the mature hepatocytes, a characteristic of the (post natal) canal of Hering (6). These findings suggested that the intralobular ducts (in hepatic cords) and their cyst complexes did not serve as breeding sources for the Meyenburg complexes or the portal cysts. Either of the latter two were neither observed by Melnick nor by us to be lined in any parts by mature hepatocytes.

The channels, projecting from the cystic canals of Hering into the portal septa, were proximally traced and found to be communicating with the bile ducts, hence concluded to be the inter-lobular bile ductules. In the present study, majority of the portal cysts and Meyenburg complexes were associated with these inter-lobular ductules. Earlier, Caroli (10) has referred to the type of the polycystic liver with the portal cysts communicating with the functioning biliary apparatus. These findings contradict the view of Meyenburg (3) that the aberrant, involuting (foetal) intra-lobular bile ducts were solely responsible for the polycystic liver and
Meyenburg complexes. In the process of development, the biliary system initially comprises of solid cords of cells which later canalise (8). An abnormality inherent in the process of canalization may lead to formation of the stenotic and weak biliary channels, liable to ectatic dilatations. This view is in conformity with the observations of Spitz (11) on the cystic dilatation of bile ducts.

The involvement of bile ducts in adult polycystic liver has been reported in one case, by Gebriel (4). He proposed that fusiform dilatations of the bile ducts in his patient could be either due to pressure on the ducts exerted by the enlarging cysts or due to congenital bile duct ectasia. In the present case cystic segments of the ducts were in series alternate with clearly defined stenotic segments, and therefore, could not be due to cyst pressure. That it was an intrinsic anomaly of the bile ducts was also supported by the findings that some of the bile ducts were involved along with their ductular and canalicular tributaries in their entire lengths while others were spared. These findings further supported and established the view of congenital biliary ectasia in the pathogenesis of adult polycystic liver and Meyenburg complexes.

Adult polycystic liver is an autosomal dominant state. The extent of the cystic involvement varies from one person to another and from generation to generation which could result from variance of penetrance in the transmission of the disease (4). The case documented here is the first example of a wide spectrum polycystic liver, extending from the bile ducts up to the bile canaliculi. The Meyenburg complexes were related to the ectatic interlobular ductules and the bile ducts.

References:


5) Adams, C. M., Danks, D. M., Campbell, P. E.: Comments upon the classification of infantile polycystic disease of the liver and kidney, based upon three dimensional reconstruction of the liver. Journal of Medical Genetics, 11: 234, 1974.


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