Nephropath Teaching Point 5-Renal Dysplasia

Last nephropath teaching point question pertained to histological features most consistently seen in biopsies featuring renal dysplasia. Majority of participants (50%) rightly agreed that presence of cartilage is not an invariable occurrence in these biopsies. In fact, metaplastic cartilage is seen in only about 1/3 rd of biopsies with renal dysplasia (though when present, diagnosis of dysplasia is certain). Other features mentioned in the question viz. disorganized tubular/ductal structures and smooth muscle collars around primitive tubules are diagnostic hallmarks and glomerulocystic change is a fairly common occurrence.

Renal dysplasia is strictly a histological diagnosis and a major cause of childhood renal failure and ESRD. It includes a constellation of clinical conditions where in spite of normal initiation of development, further maturation of glomeruli and collecting system does not occur. Renal dysplasia has been variously described based on morphological gross characteristics as unilateral or bilateral multicystic, hypoplastic, hypodysplastic or segmental dysplasia; terms which represent varied phenotypes of the same pathologic process. Pathogenesis of renal dysplasia has been a subject of intensive research and it is now that we are gaining new insights into the pathophysiological processes. Classically the two theories of primary ureteric bud failure and obstruction to urine outflow have been propounded, however recent studies have documented deregulation of gene expression correlating with disturbed cell turnover and maturation. Mutations of nephrogenesis genes have been defined in multiorgan dysmorphic disorders in which renal dysplasia can feature, including Fraser syndrome (FRAS1, FREM1 & GRIP 1), renal cysts and diabetes (HNF 1b), Kallmann syndrome (KAL 1) etc. A recent study suggests the role of Matrilysin (MMP-7) Inhibition of BMP-7 induced renal tubular branching as an important phenomenon in pathogenesis of renal dysplasia. Two excellent articles on classification of renal cystic diseases (Bonsib) and the evolving concepts in renal dyspalsia (Woolf et. al) are a must read.

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