IS2-03 Multicystic adenomatoid pancreatic hamartoma in a child: Case report and literature review

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Case: A 14-month-old boy was referred for abdominal distension. He had leukocytosis, elevated serum C-reactive protein and pancreatic enzymes (amylase: 242 IU/L; lipase: 1648 IU/L), and normal tumor markers. Diagnostic imaging initially identified a huge multicystic lesion that changed to large and small cysts with a solid component over 2 months. Pancreatic blastoma, pancreatic pseudocyst, even lymphangioma were suspected. At laparotomy, a huge multicystic/partially solid tumor originating from the tail of the pancreas was identified and resected completely without pancreatectomy. On histopathology, the cyst wall was lined with pancreatic cuboidal/tall columnar epithelium with immature pancreatic mesenchyme in the stroma, suggestive of multicystic adenomatoid pancreatic hamartoma (MAPH).

Literature review: MAPH is extremely rare. Only 17 cases (including this case) have been reported in the English literature: 4 in children and all less than 3 years old. All 4 pediatric cases presented with abdominal distension; one was hypoglycemic. Radiographic findings were non-specific. Tumors were in the head of the pancreas in 2, tail of the pancreas in 1, and diffuse in 1. Treatment was local resection in 2 and pancreateicoduodenectomy in 2. The hypoglycemic case had pancreaticoduodenectomy but died.

Conclusion: This is the first report of MAPH in a child displaying radiographic changes over time. Although rare, MAPH should be considered in the differential diagnosis of cystic pancreatic tumor in children.