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Segmental Arterial Mediolysis Presenting as a Pancreatic Mass in a Pediatric Patient: A Case Report

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Abstract: We describe a case of segmental arterial mediolysis (SAM) in a 2-year-old male who presented with symptoms of acute pancreatitis. SAM is a vascular entity of unknown etiology that involves medium-sized arteries in which the integrity of the vessel wall is compromised, resulting in increased susceptibility to ischemia, hemorrhage, and dissection. The clinical presentation is variable and can range from abdominal pain to more ominous findings of abdominal hemorrhage or organ infarction. This entity should be considered in the correct clinical setting and after other vasculopathies have been excluded. We aim to bring awareness to pediatric providers given this is a rare entity with variable presentation, which could be potentially life threatening.

Key Words: segmental arterial mediolysis, acute pancreatitis, pediatrics, pancreatic mass, vascular lesion, vasculopathy

Segmental arterial mediolysis (SAM) is a vascular entity of unknown etiology that involves the integrity of the arterial wall of medium-sized vessels, resulting in increased susceptibility to ischemia, hemorrhage, and dissection (1). Most cases have been reported in the adult population and only two cases have been reported in children, one in a preterm infant and another involving an intracranial presentation in a 14-year-old patient (2,3). We present a case of SAM in a 2-year-old male.

CASE PRESENTATION

A 2-year-old boy was hospitalized at an outside institution for acute pancreatitis (Lipase: 2754 U/L) after presenting with 2 days of fever, anorexia, abdominal pain, and vomiting. Abdominal magnetic resonance imaging revealed a 3-cm lesion within the pancreatic head (Figure 1). The main pancreatic duct was mildly dilated, and the lesion was connected to the main pancreatic duct. Abnormal ductal drainage of the pancreas was also identified, and the ductal anatomy was distorted by the mass. The remainder of the pancreas was normal without additional findings suggestive of pancreatitis. The patient was managed supportively with discharge diagnosis of acute pancreatitis with an acute necrotic collection. He followed up with his primary gastroenterologist at an outside institution.

The patient was readmitted 6 months later with another episode of acute pancreatitis (lipase: 242 U/L). CBC was notable for elevated WBC (15.9 TH/cm^2) with neutrophil predominance, platelets (120 k/μL), hemoglobin (10.7 g/dL), and elevated CRP (10 mg/dL). Magnetic resonance cholangiopancreatography (MRCP) showed enlargement of the pancreatic lesion with compression of the portal vein. There were suggestions of portal hypertension including multiple varices along the stomach and splenomegaly. The differential diagnosis included: duplication cyst, pseudocyst, pancreatic tumor, congenital pancreatic cyst, vascular changes secondary to underlying connective tissue disease or vasculitis. The patient was transferred to our institution for higher level of care.

The patient underwent explorative laparotomy for pancreatic tumor resection with pancreatoduodenectomy, pancreaticojejunostomy, and en-bloc resection of portal vein without reconstruction. The intraoperative findings included an 8 cm poorly circumscribed pancreatic head mass with compression of the portal vein. Portal venous collateralization was identified within the lesser sac with dilation of the coronary vein and regional lymphadenopathy.

Histological findings included evidence of foci of organizing hemorrhage surrounded by fibrosis and bland myofibroblastic proliferation admixed with dense collagen fibers. Adjacent to the area of hemorrhage, there was evidence of an abnormal artery with intimal hyperplasia. A Movat stain revealed segmental loss of the internal elastic lamina and tunica media indicative of aneurysm formation (Figure 2). There was no evidence of malignancy or abnormal lymph nodes. Additional vascular imaging studies did not reveal any other vascular involvement. The

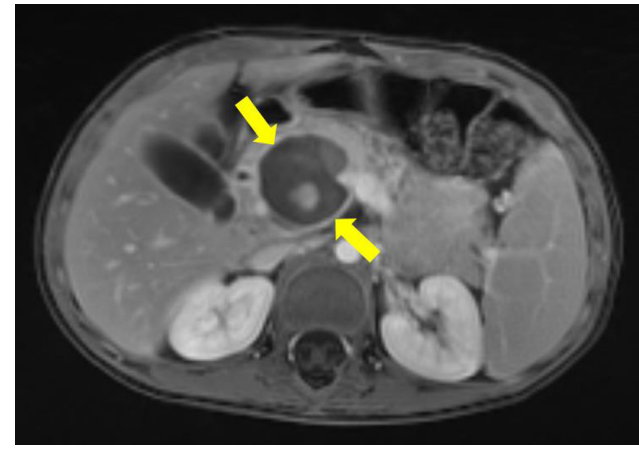

FIGURE 1. MRI abdomen T1 VIBE postcontrast axial image. No flow or enhancement is seen within a 3-cm lesion within the pancreatic head (arrows). This image is unchanged compared with the precontrast VIBE sequence. MRI = magnetic resonance imaging; VIBE = volumetric interpolated breath-hold examination.

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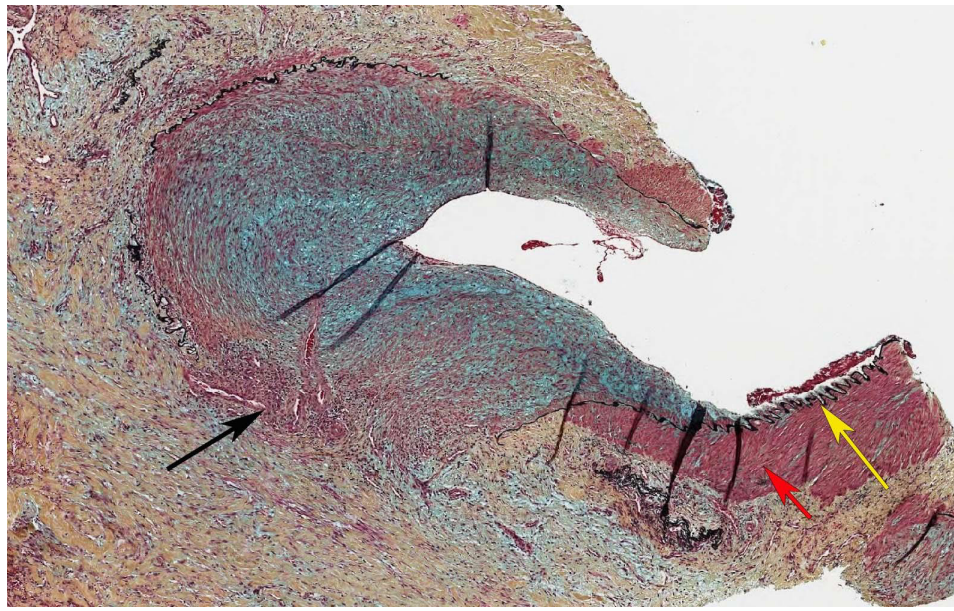

FIGURE 2. Segmental loss of the elastic lamina (black arrow) and tunica media (red arrow) within an abnormal artery (Movat Stain). The yellow arrow highlights internal elastic lamina.

overall findings were most indicative of a ruptured aneurysm secondary to a noninflammatory vasculopathy, most consistent with SAM. After resection, 3 subsequent computed tomography angiograms performed over a 2-year period did not show recurrence of SAM.

DISCUSSION

SAM is a rare vasculopathy that is noninflammatory and nonatherosclerotic in nature (4). The incidence of SAM is unknown and most of the knowledge about the disease is derived from existing case reports. The first case of SAM was described by Slavin and colleagues in 1976. This group found that SAM typically occurred in the abdominal splanchnic arteries in the elderly, but could also involve the coronary arteries (5). Since 1976, more than 100 cases have been reported in the adult literature, but SAM is uncommon in pediatrics. To our knowledge only 2 pediatric cases have been reported. Eifinger et al reported a case of a preterm infant with SAM diagnosed postmortem, localized to the placental, umbilical, splenic, and cerebral vasculature (2). Cayrol et al described a case of cerebral SAM in a 14-year-old male identified postmortem following a right parietal intracranial hemorrhage (3). Our case adds to the pediatric literature and is unique in that the diagnosis was identified during the early phase of clinical presentation.

The etiology is not entirely understood, but it is hypothesized that SAM results from vasospasm due to high levels of catecholamines and other vasoactive substances (6). Vasospasm may be triggered by hypoxia, hypotension, or sepsis, which results in an inappropriate response of the vascular endothelium. In a study evaluating stillborn and newborn infants, mediolytic necrosis was identified in the outer part of the coronary arteries, which were thought to be a result of perinatal hypoxia (7). Our patient had no history suggestive of perinatal hypoxia or abdominal trauma.

Presentation of SAM is variable ranging from asymptomatic or abdominal pain to dramatic life-threatening hemorrhage. Diagnosis of SAM is based on a combination of clinical features and histopathology findings. The 2 histologic stages in SAM are the injurious and reparative phases (8,9). Characteristic histologic finding in the injurious phase involves lytic degeneration of the tunica media leading to tearing or separation from the adventitia (4,8). Transmural mediolysis can result in dissecting hematomas and aneurysm formation (10). The reparative phase involves tissue overgrowth of intima leading to stenosis (8).

Although histology is considered the gold standard for the diagnosis of SAM, visceral angiography plays a key role in identifying the vasculature involved and patterns of involvement such as stenosis, dissections, or aneurysms. Common angiographic findings include aneurysm, beading or web formation, occlusion, and wall thickening (11). Although any vessel may be affected, celiac artery and the superior mesenteric artery branches are the most commonly involved (12). SAM can rapidly evolve and serial angiography may be required (13). Treatment strategies include surgical management or arterial embolization depending on the clinical presentation and vascular involvement. In our patient, a pancreatic mass was initially identified on MRCP and the final diagnosis of SAM was made based on histopathology findings as described above.

The challenging task for the physician is to distinguish SAM from other vascular disorders such as of fibromuscular dysplasia (FMD) (1,14). FMD differs from SAM in that it usually affects middle-aged females, smokers, and often causes renal artery stenosis (1,14). Arterial stenoses are the hallmark finding in FMD compared with aneurysms and dissections seen in SAM (14).

Other differential diagnoses to be considered in this context include neurofibromatosis, vasculopathies, connective tissue disorders such as Marfan's Syndrome and Ehler's-Danlos. The clinical presentation (lack of musculoskeletal findings or systemic inflammation), histological features (lack of vascular inflammation), type (small-, medium-, or large-sized vessels), or location of vascular involvement (visceral mesenteric or renal or coronary or intracranial) and laboratory findings can help differentiate these from SAM.

In conclusion, SAM is a vascular entity in which the integrity of the vessel wall is compromised, resulting in increased susceptibility to ischemia, hemorrhage, and dissection (1). SAM should be considered in the setting of distinctive combination of clinical features, angiography findings, and histopathology after excluding other vasculopathies. Evaluation and management of SAM should be multidisciplinary as SAM could be potentially life threatening.

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