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EDUCATION AND IMAGING

Hepatobiliary and Pancreatic: Pancreatic vascular malformations in hereditary hemorrhagic telangiectasia

A woman, aged 70, with hereditary hemorrhagic telangiectasia had investigations because of an 18-month history of intermittent pain in the right upper quadrant of her abdomen that radiated into the back. An ultrasound study and computed tomography (CT) scan revealed a small vascular mass at the junction of the head and body of the pancreas that raised the possibility of an islet cell tumor. A repeat enhanced CT scan after 8 months showed vascular lesions in the head (Figure 1 above) and body (Figure 1 below) of the pancreas. A subsequent magnetic resonance imaging (MRI) scan showed lesions that were thought to be atypical for arteriovenous malformations and more consistent with a pancreatic neoplasm. Various tumor markers including CA19.9 were within the reference range. She was referred for endoscopic ultrasound (EUS) with a view to fine needle aspiration. Three hypoechoic lesions, 8–10 mm in diameter, were noted in the head, neck and body of the pancreas. Color Doppler examination of all lesions showed a densely vascular pattern that filled the whole lesion (Figure 2). As she was known to have hereditary hemorrhagic telangiectasia, the lesions were diagnosed as vascular malformations and biopsies were not performed. She remains clinically stable after 2-years of follow-up. Neither the size nor number of pancreatic lesions has changed on repeat CT and EUS.

Hereditary hemorrhagic telangiectasia, otherwise known as Osler-Weber-Rendu disease, is an autosomal dominant disorder characterized by vascular abnormalities on mucosal surfaces and within internal organs. The prevalence of the disease is approximately 1 in 5,000–8,000 people. Most but not all patients have recurrent bleeding from the nasal mucosa. Up to one-third of patients have bleeding from telangiectasia in the gastrointestinal tract, particularly from the stomach and duodenum. Arteriovenous malformations also occur in the lungs (10%), brain (5–10%) and liver (5–20%). Arteriovenous malformations in other organs including the pancreas are rare. In the patient described above, the diagnosis of pancreatic arteriovenous malformations was supported by EUS with Doppler and CT scans but was less certain with MRI. However, EUS with Doppler showed typical vascular lesions that did not require histological evaluation. This conservative approach has been supported by follow-up studies.

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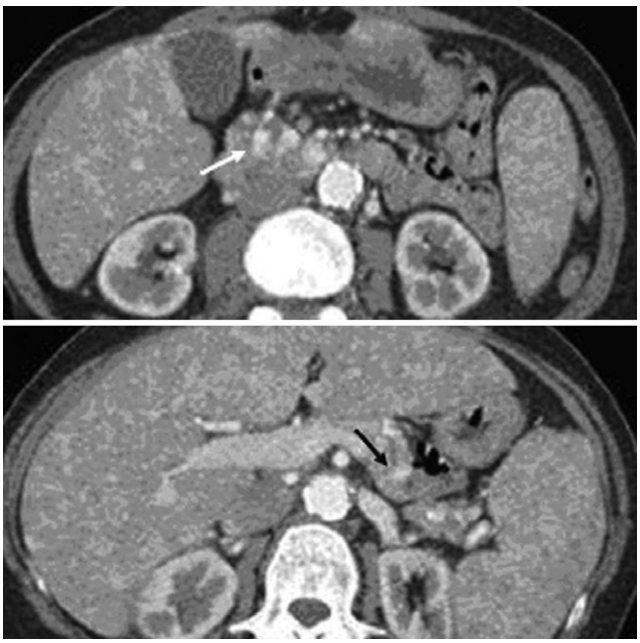

Figure 1

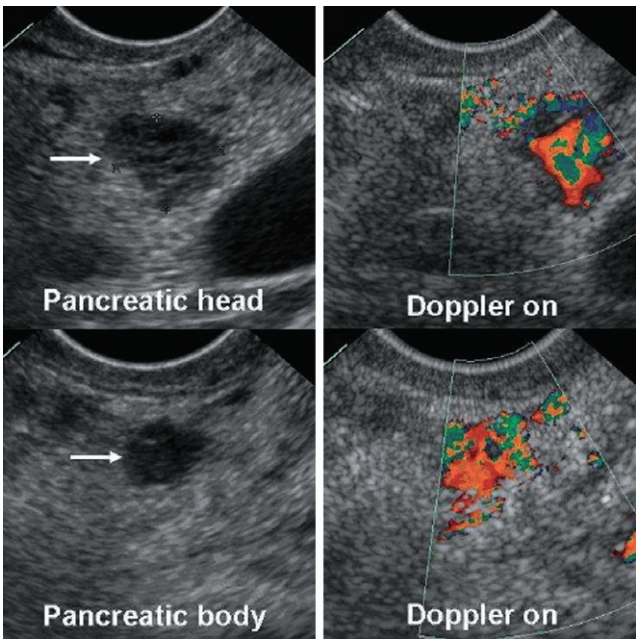

Figure 2