

CLINICAL QUIZ

Splenic Hilar Mass With Pain, Diarrhoea and Sweating: Can It Be Malignant?

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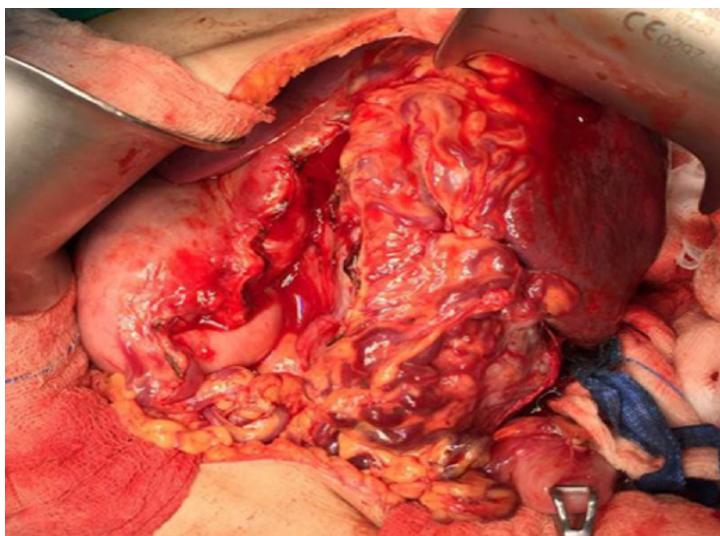


Figure 1: A hard nodular mass at the hilum of spleen with dilated tortuous vessel

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Figure 2: macroscopic picture of splenectomy and distal pancreatectomy specimen

QUESTION

A 37-year-old lady with no known comorbidity complained of left hypochondrial pain for 6 months duration. It was associated with loss of weight and appetite with occasional carcinoid symptoms such as diarrhoea and sweating. She had no blood transfusion before. Clinically, she has no features of thalassemia and the abdomen was soft with a palpable vague mass on the left hypochondrial region. A computed tomography (CT) of the abdomen revealed a soft tissue mass measuring 8.9 x 10.3 x 9.0 cm in the enlarged spleen. A laparotomy was decided and the intraoperative findings are as in Figure 1 and 2. Spot the diagnosis and provide the outline of management.

ANSWER

The diagnosis is non-functional distal pancreatic neuroendocrine tumour (PNET), based on the clinical presentations. It is evidenced by the presence of carcinoid symptoms in the background of healthy individual. It can easily mimic splenic angiosarcoma intraoperatively. PNET originates from diffuse neuroendocrine or islet cells of the pancreas. It only comprises of 2% of total number of cases making it a surgical rarity. According to World Health Organization (WHO) 2010, PNET is categorized into functional and non-functional neuroendocrine neoplasm based on the existence or non-existence of symptoms caused by hormonal

hypersecretion. Among the involved hormones in the functional PNET include insulin, gastrin, glucagon, somatostatin, and vasoactive intestinal peptide in which insulinoma exhibits the most common aetiology. For the non-functional PNET, local compression and metastatic lesions are most likely causes to the problem. In a symptomatic individual, the main complaints include abdominal pain, weight loss and nausea which are non-specific. Hence, serum chromogranin A can be an adjunct investigation to the diagnosis. Undeniably, the role of imaging such as the conventional ultrasonography, computed tomography, and magnetic resonance imaging is helpful, but by performing gallium-68 labelled studies and Octreotide analog studies in combination with positron emission tomography are excellent. In most cases, surgical resection remains the opted modality of choice. However, in a small, incidentally discovered, or asymptomatic non-functional PNET, vigorous observation can be chosen for conservative treatment.