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Inflammatory pseudotumour of the pancreas in a child

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Abstract We describe a 4-year-old girl with an inflammatory pseudotumour of the pancreas, which was preceded by varicella-zoster infection. Inflammatory pseudotumour may involve a variety of tissues, the lungs and liver being typical sites of predilection. Imaging and laboratory tests are nonspecific, and for this reason the diagnosis of inflammatory pseudotumour is rarely made prior to surgery. These benign but locally aggressive masses simulate malignancy in the majority of cases. Inflammatory pseudotumour should, therefore, be considered when a mass arises in an unusual location in the paediatric age group.

Introduction

Inflammatory pseudotumour (IPT) is an uncommon condition that has more recently been classified as an inflammatory myofibroblastic tumour. This condition produces a benign mass comprising inflammatory cells, fibrous stroma and spindle cells, and has been described in a variety of locations, the pancreas being an infrequent site. Preoperative diagnosis is difficult and confusion may arise when imaging shows a mass and gives an erroneous impression of malignancy. The aetiology of this condition is uncertain, but hypotheses include immunological causes and an inflammatory response to insults such as infection or trauma. A 4-year-old girl with a pancreatic (IPT) and prodromal varicella-zoster infection is presented. This case lends support to the hypothesis that IPTs represent a response to infection.

Case report

A 4-year-old girl was admitted to hospital with a 4-week history of malaise, lethargy and a vesicular skin rash. The rash had resolved only to recur some 2 weeks before admission. There had been diarrhoea for the past 3 days and skin discolouration, in keeping with jaundice, developed 24 h prior to admission. On examination, pallor and jaundice were present. There was a vesicular rash on the trunk and extremities. A soft, smooth liver edge was palpable 3 cm below the right costal margin and the provisional clinical diagnosis was of viral infection.

Viral culture of the skin lesions revealed varicella-zoster virus. Cultures for CMV and EBV were negative, as was faecal culture. Hepatitis A, B and C, and herpes simplex serology were negative. Liver function tests showed a predominantly biliary obstructive picture. Upper abdominal US indicated a mass in the pancreatic head and CT was performed for further evaluation. CT revealed an enhancing mixed-attenuation mass within the pancreatic head (Fig. 1) with mild dilatation of the pancreatic and intrahepatic bile ducts. CT-guided fine-needle aspiration was performed and was suggestive of pancreatoblastoma or an epithelial neoplasm, possibly of neuroendocrine or islet cell origin. Following transfer to a

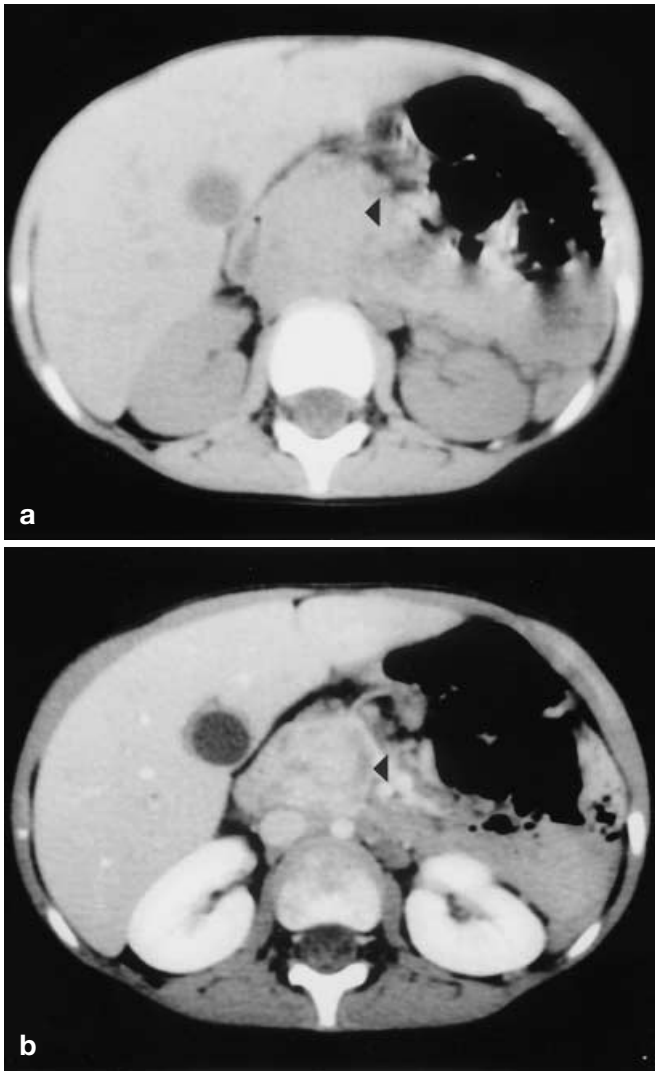

Fig. 1 a, b Axial CT through the upper abdomen. **a** Unenhanced scan revealing a 3-cm diameter mass (*arrowhead*) in relation to the head of the pancreas. The mass was of similar attenuation to muscle and did not contain macroscopic calcification. **b** Contrast-enhanced portal venous phase shows heterogeneous enhancement (*arrowhead*) and close proximity of the mass to the aorta and inferior vena cava. Biliary dilatation secondary to the mass was present on more cranial slices

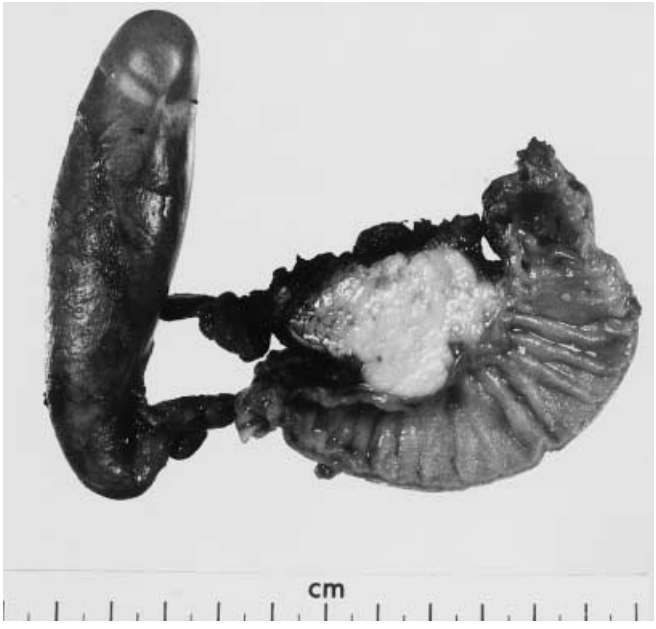

Fig. 2 The resected specimen showing duodenum, head of pancreas with pseudotumour and gall bladder

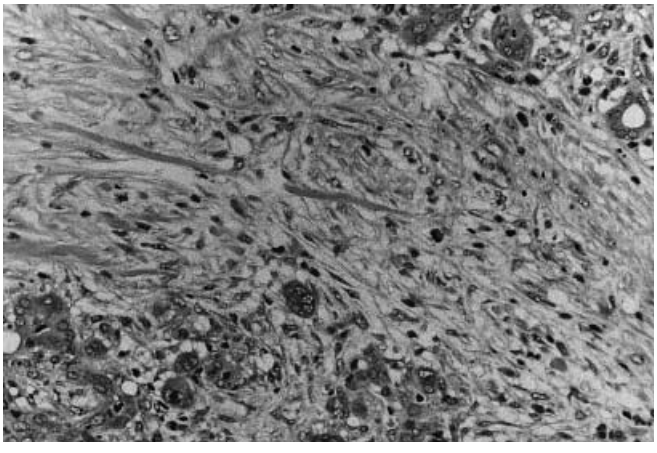

Fig. 3 Histology shows proliferating fibrous tissue with residual pancreatic acinar cells. (H&E stain; original magnification, × 80)

paediatric tertiary referral centre, a Whipple's procedure was performed because of biliary and pancreatic duct obstruction and suspected malignancy. At operation a mass confined to the pancreatic head was confirmed, and there was no evidence of tumour spread.

The surgical specimen consisted of gall bladder, duodenum and head of the pancreas. There was a firm 30 × 30-mm mass with a yellow-white appearance on cut section (Fig. 2). The common bile duct was unable to be probed within the mass. Microscopically, the lesion was characterised by fibroblastic proliferation with an acute-on-chronic inflammatory infiltrate in which eosinophils were conspicuous (Fig. 3). Viral inclusions were not identified. The appearances were considered to be those of an IPT. Other pathological opinions were sought, and it was generally agreed that the inciting process was probably infectious and that varicella was the likely cause.

The patient made a rapid and uncomplicated recovery, was on TPN for 6 days and was feeding orally after 12 days. Over the next 4 years she continued to thrive, her growth virtually matching that of her identical twin.

Discussion

IPT is an uncommon benign condition that frequently simulates malignancy because it forms a mass. The com-

monest site of origin is the lung, over 110 cases having been reported in this location [1]. The liver is also relatively frequently involved [1], but other sites such as the stomach [2], spleen [3], bladder [4], kidney [5], maxillary sinuses [6], heart [7], parapharyngeal space [8], retrorectal space [9] and peripheral nerve [10] have also been recorded. IPT has a number of synonyms, including inflammatory myofibroblastic tumour [11], plasma cell granuloma and xanthofibroma. The condition is observed in patients of widely differing ages and, although locally aggressive, it does not metastasise.

To our knowledge, only 11 cases of pancreatic IPT have previously been described [12–17]. The presentation of pancreatic IPT varies, constitutional symptoms, a mass, pain and jaundice having been recorded [12]. Although microcytic anaemia has been noted in some patients with IPT, this was not present in our case. Laboratory findings are nonspecific, but may include hypergammaglobulinaemia and a high erythrocyte sedimentation rate. Imaging typically reveals a mass and the usual presurgical diagnosis is malignancy.

CT appearances are variable. The mass may be hypodense or isodense to muscle on pre-contrast scans and calcification has been observed within inflammatory pseudotumours of the pancreas [14], stomach [2] and liver [18]. Contrast enhancement is usually not pronounced, but a variety of patterns has been noted. These include delayed peripheral, heterogeneous, homogeneous and no enhancement.

Histology is characteristic and shows a proliferation of plasma cells, other mononuclear and inflammatory cells, a fibrous stroma and spindle-shaped mesenchymal cells [12]. In general, the prognosis is excellent after surgical resection has been performed, local recurrence being an infrequent complication [17].

The origin of IPT is unclear, an inflammatory response to infection, trauma or surgery and immunological causes having been proposed. Although latent Epstein-Barr virus (EBV) genome has been observed in IPT of the liver and spleen [19], a previous report of localised lymphoplasmacellular pancreatitis forming a pancreatic IPT did not confirm the presence of latent EBV [16]. Varicella-zoster infection has not been previously reported in association with pancreatic IPT. Although events in this case suggest development of the pseudotumour in response to pancreatic varicella-zoster infection, this could not be confirmed.

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