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Case Report

Solid pseudo-papillary tumour (SPT) of the pancreas, an unusual cause for multi-nutritional deficiency anemia

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Introduction

Pseudo-papillary neoplasms of the pancreas are rare pancreatic tumours that account for up to 1-3% of all pancreatic tumours. They have a relatively low potential for malignant transformation [1]. Most of these tumours are found in females in their 2nd and 3rd decade of life. Definitive treatment is complete surgical resection, and this usually has a good prognosis [2]. We describe an 11-year-old girl with symptoms of anaemia for 2 months, later diagnosed to have a solid pseudo-papillary tumour of the pancreas.

Case Presentation

A previously healthy, 11-year-old girl was transferred from a local hospital to Lady Ridgeway Hospital for further evaluation of severe anemia. She was born to non-consanguineous parents from a middle-class socio-economic background. She had developed gradual onset of exertion-induced dyspnoea. She had easy fatiguability and palpitations on exertion. However, she could attend to her daily routine activities. There were no features of heart failure nor a history of febrile illness, bleeding manifestations or jaundice. She had lost 2 kg over two months but denied evening pyrexia, anorexia, or bone pain. She did not have abdominal pain, fullness, or vomiting. Her bowel habits were normal and there was no food intolerance. Her diet comprised rice, green leaves, vegetables, and food of animal origin, like eggs, fish and meat, in adequate quantity. She did not have a family history of malignancy.

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As the symptoms were getting worse, she was admitted to the local hospital. There she was found to have a low haemoglobin (Hb) of 4.6g/dl that required a blood transfusion and evaluation for anaemia. Later, she was transferred to the Lady Ridgeway Hospital for Children following a contrast enhanced CT scan of the abdomen that revealed a mass lesion over the pancreas.

On admission, she was pale. Her anthropometry revealed a weight of 40kg (75^{th} centile), height of 140 cm (25^{th} centile) with BMI of 21.3kg/m^2 (90^{th} centile). She was rational and conscious. Her respiratory rate was 20 cycles/min, heart rate 110bpm and blood pressure 116/65mmHg. The apex was not deviated and no crepitations were heard over the lung bases. Her abdomen was soft and non-tender and there was no hepato-splenomegaly or palpable masses. Her neurological examination including gait and fundoscopy was normal.

Table 1 shows the summary of investigations done at the tertiary center. Her contrast enhanced computer tomography (CECT) images of the abdomen are shown in Figure 1.

Investigations	**Values**	**Normal Range**
WBC	$12.8*10^3$/ml	$4.5 -10.0^{*10}3$/ml
N/L	75%/21%	
Hb	6.5g/dl	12.1-15.1 g/dL
MCV	87fl	80-95 fl
MCH	24pg/cell	27-31 pg/cell
MCHC	28g/dl	32-36 gm/dL
Platelet	$466*10^3$	$150-400*10^3$
Retic count	3.4%	0.5%-2.5%
LDH	146.6 U/l	140-280 U/l
ALT	29IU/l	0-45IU/l
AST	20IU//	0-35IU/l
Albumin	4.2g/dL	3.5 -5.5g/dL
CRP	2mg/l	0.8 -3.0mg/l
ESR	45mm/hour	1-13mm/hour
Serum Na	137 mmol/l	136-144mmol/l
Serum K	4.3mmol/l	3.7-5.1mmol/l
Serum creatinine	40micromol/l	52.2 - 91.9 micromol/l)
Serum Ferritin	11.74microgram/l	11-307microgram/l
Vitamin B 12	339pg/ml	200-1100pg/ml
Stool for occult blood	negative	
Ca 19.9	24	0-37

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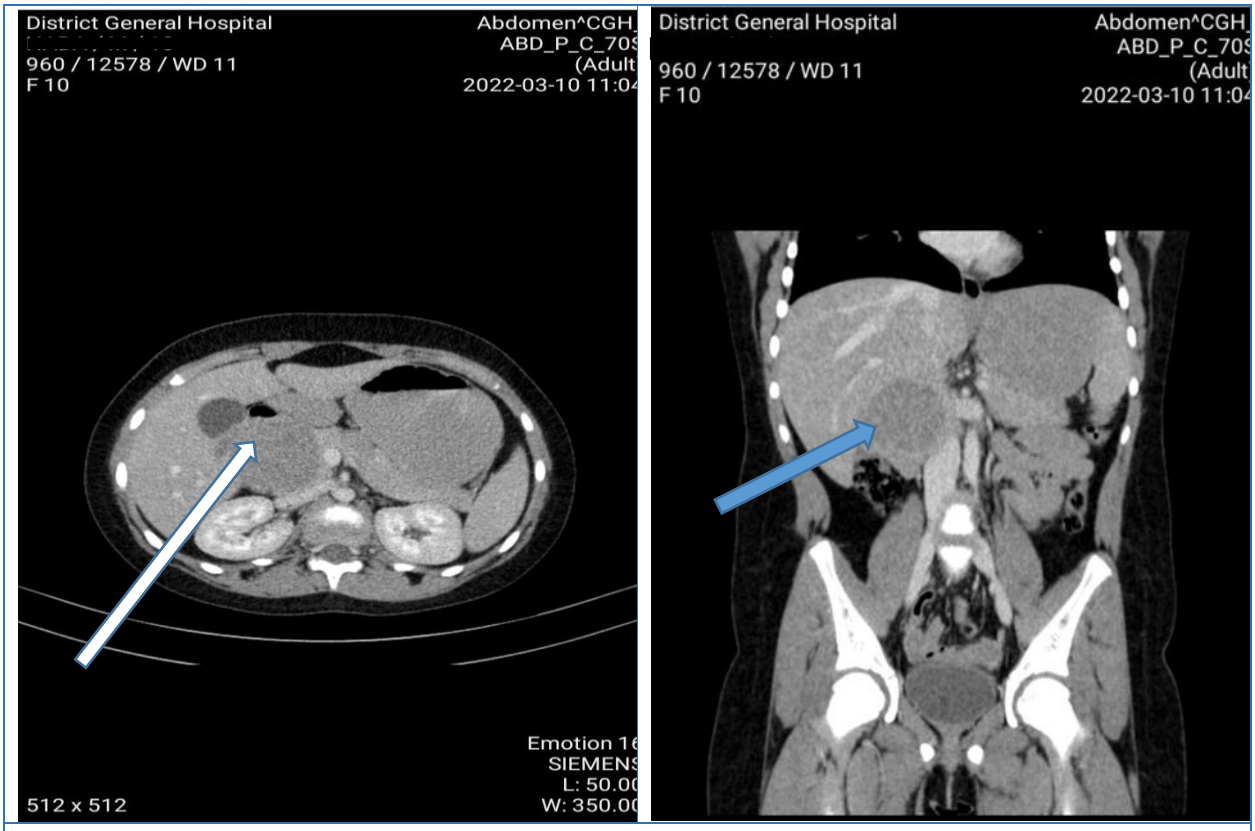

Figure 1: There is a well-defined hypodense homogenously enhancing solid mass (shown by an arrow) in the pancreatic head region. It measures 6.7*6.1*5.9cm in size. Appearances are in favour of a pancreatic head neoplasm.

Bone marrow aspirate and trephine biopsy revealed subpopulations of hypochromic, microcytic red blood cells, hypersegmented neutrophils and a few megaloblasts compatible with multi-nutritional deficient anaemia. However, there was no evidence of marrow infiltration by a heamatological malignancy.

Subsequently, she started on a trial of Vitamin B 12 1000mcg with folic acid. She was referred to a paediatric surgeon. An ultrasound guided FNAC revealed a solid pseudo papillary tumour of pancreas. She later underwent Whipple's procedure (pancreatic-duodenectomy). The histology of the resected tumour revealed papillae with a fibrous core, lined by columnar cells with regular nuclei with reversed polarity suggestive of a solid pseudo-papillary tumour of the pancreas. Her perioperative period was uneventful and after two weeks of hospital stay, she was referred to an oncologist and nutritionist.

Discussion

Solid pseudo-papillary tumours (SPT) of the pancreas are rare in children and their pathogenesis is not yet fully understood. The site of tumours is different in children and adults [3]. In children, they are found in the head of pancreas, whereas in adults they are

found in the tail and body [2]. These tumours are found in head, body, and tail of pancreas and sometimes in the retroperitoneum, liver, lung, omentum and duodenum outside the pancreas [3,4].

Even though our index patient did not have any of these symptoms, patients can present with abdominal discomfort, indigestion, epigastric pain, nausea, vomiting, weight loss, early satiety and bloating due to compression of the adjacent stomach. They usually do not have features of pancreatic exocrine or endocrine insufficiency [3,4,5].

Risk factors for pseudopapillary tumours of the pancreas are under-studied. However, well-known risk factors for pancreatic carcinoma are dietary habits like high alcohol intake, fructose and red or processed meat intake, type 2 diabetes, obesity, stress, and smoking [6,7]. None were present in our patient.

When clinically suspected, an ultrasonogram can be arranged initially, which may show a homogenous, hypoechoic mass with a hyperechoic rim. MRI and contrast-enhanced CT are superior to ultrasonogram. [8]. CT may show an encapsulated mass with solid and cystic components with calcifications in the periphery. MRI, the preferred choice of imaging, may show an encapsulated lesion with both solid and cystic components with haemorrhage, sometimes in the absence of septation [8]. Histological evaluation by endoscopy guided biopsy and immunohistochemistry studies are helpful in making an accurate pre-operative diagnosis. [1] Tumour markers like carcinoembryonic antigen (CEA) and CA19.9 are usually normal [8].

Surgical resection is considered the definitive treatment of choice [1,2] as was successfully done in our patient. The type of surgery depends on the tumour size, site and its extension. Salvaging organs is advocated, if feasible [1]. Overall survival after surgery is excellent if complete resection is feasible, even in the presence of metastasis [1,2,5]. There are a few published cases in Sri Lanka. These publications mainly focused on pathology and diagnostic evaluation [9].

Conclusion

Anaemia in a child could be due to multiple reasons, and if history, examinations and basic investigations are not supportive of a more common cause, a sinister aetiology, such as malignancy, should be thought of. The patient being a girl and obese might have been contributory factors for developing a rare malignancy like SPT of pancreas, prompting the need for further studies and research.

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