A child with neuroblastoma who presented with acute pancreatitis

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Introduction

Neuroblastoma is the commonest extracranial solid tumour in children and accounts for 15% of all paediatric deaths due to malignancies[1,2]. It commonly presents as abdominal pain and abdominal mass. It may present with obstructive jaundice due to bile duct obstruction[3-6]. We report a case of neuroblastoma with acute pancreatitis.

Case report

An eight year old boy was admitted with a history of abdominal pain for 3 months and abdominal distension for 1 month. The abdominal pain was dull, severe, agonizing, mostly confined to the epigastric region, radiating to the back, aggravated after taking food, relieved by leaning forward and associated with anorexia and diarrhoea. Abdominal distension was gradually worsening (Figure 1). There was no history of taking any drugs such as azathioprine, 6-mercaptopurine, methyldopa or thiazide diuretics and no history of abdominal trauma or previous viral illness. There was, no family history of pancreatitis and no contact history of tuberculosis.

Two days after admission, he developed continuous fever (highest temperature 104^0F) and non-bilious vomiting. On examination on day 3 of admission, he was fretful, febrile, had mild pallor, a pulse rate of 84/min, blood pressure of 110/70 mm Hg, a respiratory rate of 34/min and oxygen saturation (SPO2) of 97% in room air. Anthropometrically the Wight for Height Z-score (WHZ) was -3.49 and the Height for Age Z-score (HAZ) was -2.7 with age-appropriate development. Generalized lymphadenopathy was present, which was firm in consistency, non-tender, discrete and mobile. The abdomen was distended with slit like umbilicus. An ill-defined mass was present in the umbilical and epigastric regions, firm in consistency with an irregular surface, not attached to skin or underlying structures. There was no organomegaly. Ascites was present. Other system examination was normal.

Figure 1: ***Abdominal distension***

Laboratory and radiological investigations are summarised in Table 1. There was microcytic hypochromic anaemia. Serum lactate dehydrogenase, serum lipase and serum alanine aminotransferase were markedly elevated. The ascitic fluid had a low Serum Ascites Albumin Gradient (SAAG). Ultrasonography of abdomen showed moderate ascites, enlarged para-aortic, pre-aortic, peri-pancreatic and pelvic lymph nodes and a compressed and swollen pancreas. Computerized tomography (CT) of the abdomen showed a swollen pancreas and a mass in the pre and para-aortic region with moderate ascites (Figures 2 and 3). An ultrasound guided biopsy from the abdominal mass was done. Histopathology showed a small round blue cell tumour (Figure 4) and immunohistochemistry showed CD 56 positive in tumour cells compatible with neuroblastoma. After evaluating the clinical data, physical findings, and investigation results, the case was finally diagnosed as neuroblastoma presenting with acute pancreatitis. The patient was referred to paediatric haematology and oncology for protocol-based chemotherapy.

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Table 1: ***Laboratory and radiological investigations***

Investigation	Results	Normal range
Complete Blood Count		
Haemoglobin (g/dL)	10.5	13-17
ESR (mm in 1st hr)	45	0-10
White blood cell count (/cu mm)	10,300	4,500-11,000
Neutrophils (%)	55	40-80
Lymphocytes (%)	36	20-40
Platelet count (/cu mm)	300,000	150,000-400,000
Liver function tests		
Serum albumin (g/L)	42	35-50
Prothrombin time (sec)	10.05	12-16
International normalized ratio	0.88	<1.4
Alanine aminotransferase (u/L)	282	35-50
Serum lipase (u/L)	1660	0-160
24 hours urinary VMA (mg /day)	6.5	5 – 15
Serum lactate dehydrogenase (u/L)	2023	140-250
Serum creatinine (mg/dl)	0.5	0.9-1.3
X-ray chest	Normal	
Mantoux test	Negative	
Ascitic fluid study		
Total white blood cell count (/cu mm)	400	<250
Neutrophils (%)	15	
Lymphocytes (%)	85	
Lactic dehydrogenase (u/L)	900	400
Ascites fluid albumin (g/dL)	3.4	0.3-4.0
Adenosine deaminase (u/l)	3.5	Up to 15
Ascitic fluid amylase (u/l)	143	50% 0f serum level
SAAG (g/dl)	0.8	
Ultrasonography of abdomen	Moderate ascites, Enlarged lymph node- Para -aortic, pre-aortic, peri -pancreatic & pelvic. Pancreas is compressed and swollen.	
Computerized tomography of abdomen (Figures 2 and 3)	Pancreas was swollen, a mass in pre and para-aortic region with moderate ascites	
Histopathology (Figure 4)	Small round blue cell tumour. Tumour cell have dark round to oval nuclei and small amount of cytoplasm. A few perivascular **pseudo-rosettes** are present.	
Immunohistochemistry	CD 56 positive in tumour cells compatible with neuroblastoma.	

ESR: erythrocyte sedimentation rate, VMA: vanillyl mandelic acid, SAAG: serum ascites albumin gradient

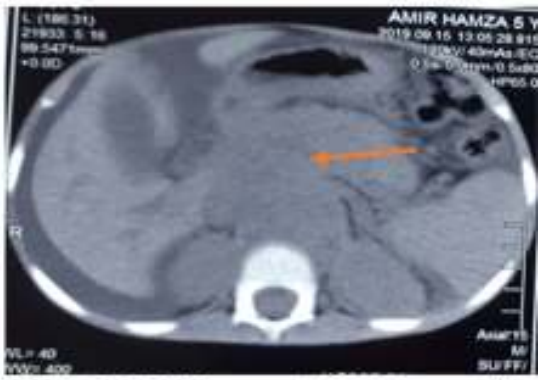

Figure 2: ***CT abdomen. Arrow shows swollen pancreas along with pre-aortic lymph node***

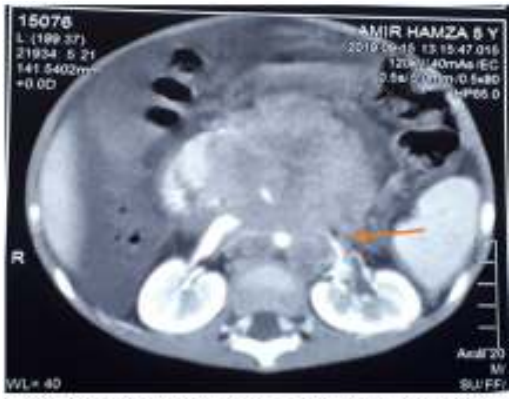

Figure 3: ***CT abdomen. Arrow shows a mass which is compressing renal vessels***

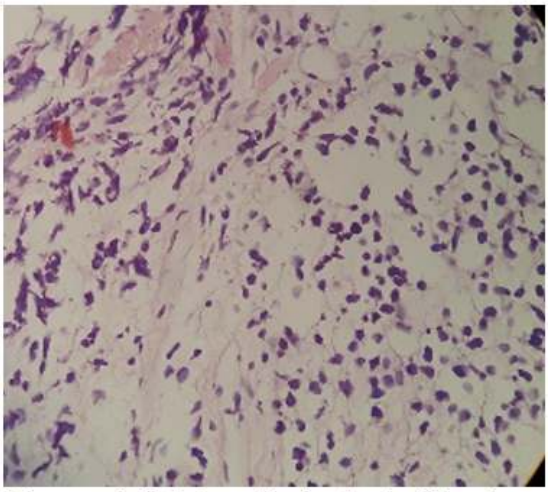

Figure 4: *Histopathological slide show small round blue cells*

Discussion

Around 90% of children with neuroblastoma are diagnosed before 5 years of age[7]. Our patient was 8 years old when diagnosed. About 60% of primary neuroblastomas arise in the abdomen and commonly present with fixed and firm abdominal mass, abdominal pain, protein losing enteropathy, ascites and weight loss[8]. Hepatobiliary obstruction is a rare complication of neuroblastoma and pancreatic ductal obstruction was reported in one case[9]. Our patient had pancreatitis which is a rare presentation of neuroblastoma and it may have occurred due to pancreatic ductal obstruction by tumour invasion of the head of the pancreas and due to cytokines released from the tumour e.g. TNF-α and IL-β. Regional lymph node metastases are noted in 35% of patients with apparently localized neuroblastoma[8]. Our patient had generalized lymphadenopathy.

Diagnosis of acute pancreatitis is made if the patient presents with at least two of the following three manifestations: acute attack of characteristic abdominal pain, elevated levels of pancreatic leaking enzymes and findings of the pancreas detected by USG, CT or magnetic resonance imaging (MRI)[10]. Our patient had abdominal pain, elevated pancreatic enzyme and swollen pancreas on USG and CT scan. Intra-abdominal malignancy especially neuroblastoma may be a rare cause of pancreatitis. Neuroblastoma should be considered in the differential diagnosis if a child presented with abdominal mass and pancreatitis.

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