ROLE OF IMAGING IN DORSAL PANCREATIC AGENESIS WITH RETROPERITONEAL LYMPHANGIECTASIA- IN A CASE OF ACUTE ABDOMINAL PAIN

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ABSTRACT

BACKGROUND

Filariasis is a parasitic infection. It is an endemic infection seen in tropical and subtropical geographical regions of the world. Lymphatic filariasis is caused by parasites Wuchereria bancrofti, Brugia malayi and Brugia timori. It may present with lymphatic dysfunction in the form of lymphocoele, chyluria, hydrocoele, elephantiasis and groin lymphadenopathy. Retroperitoneal lymphangiectasia presenting with acute abdominal pain is a rare clinical entity and very few cases are reported worldwide. Total agenesis of the pancreas is an extremely rarest congenital anomaly[1] and is incompatible with life. Although complete dorsal pancreatic agenesis is one of the rare anomaly, partial agenesis of the dorsal pancreas is more common than ventral agenesis of pancreas.[2,3] It can be associated with other congenital anomalies like polysplenia syndrome,[1-4] liver lobulation, heterotaxia and solid-pseudopapillary tumour of pancreas.[2,4] In most of the cases it remains clinically asymptomatic, but sometimes it may present with hyperglycaemia, focal pancreatitis presenting with acute or chronic abdominal pain. Here, we are presenting a case of retroperitoneal lymphangiectasia with complete dorsal pancreatic agenesis presented with acute abdominal pain. Both complete dorsal pancreatic agenesis and retroperitoneal lymphangiectasia are the two rarest clinical entities one may come across in a day-to-day practice.

KEYWORDS

Filariasis, Retroperitoneal Lymphangiectasia, Dorsal Pancreatic Agenesis.

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BACKGROUND

Filariasis is a parasitic disease prevalent in tropical and subtropical geographic regions. Lymphatic filariasis is caused by Wuchereria Bancrofti, Brugia malayi and Brugia timori. It is a type of helminthiases caused by roundworms of Filarioidea type. These worms reside in the lymph nodes, lymphatic channels, clinical manifestations are related to occlusion of the lymphatics causing lymphangiectasia.[5,6]

Congenital anomalies of the pancreas are usually discovered in childhood; however, some may develop later in adult life owing to the development of clinical symptoms which is related to the anomaly itself or symptoms mimicking acquired diseases, mainly pancreatic cancer.

At fourth week of gestation a dorsal and a ventral endodermal diverticulum arises near the junction with the yolk sac. The ventral pancreas arises from the ventral diverticulum and the dorsal pancreas arises from the dorsal anlage. Agenesis of the ventral pancreas is an extremely rare anomaly, whereas dorsal pancreatic agenesis, although a rare entity is documented in 20 case reports. Due to early embryologenetic defect, there is complete or partial absence of the structures normally arising from the dorsal pancreatic bud 2.

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A 24-year-old male from Bihar presented with acute abdominal pain in the emergency followed by one episode of vomiting and history of one day fever.

On clinical examination, there is tenderness over epigastric region. No family history of diabetes and pancreatic carcinoma were present. On laboratory investigation, leucocytosis was present.

On contrast enhanced CT abdomen study revealed multiple ill-defined soft tissue density mass lesion occupying the retroperitoneum in the pre- and para-vertebral regions encasing the abdominal aorta and its branches, inferior vena cava and bilateral renal vessels. The lesions are seen extending into the pelvis predominantly along the left iliac vessels encasing them and extending into the left inguinal region and the anterior aspect of the thigh encasing the common femoral vessels.

The neck, body and tail of the pancreas are not visualised. Both main and accessory pancreatic ducts are not visualised. On ultrasound evaluation, there were multiple ill-defined cystic structures in the peri, para-aortic regions, tubular linear anechoic cystic lesion are also noted along bilateral iliac vessels. On scrotal sonography, there was hydrocele with linear echogenic structures within it and linear as well as cystic anechoic lesions suggestive of dilated lymphatic channels with curvilinear echogenic structures within it likely microfilariae (filarial dance sign).

On ultrasound guided aspiration of cystic fluid from scrotum, there are evidence of filarial parasite. On peripheral blood smear examination, there were evidence of microfilariae.

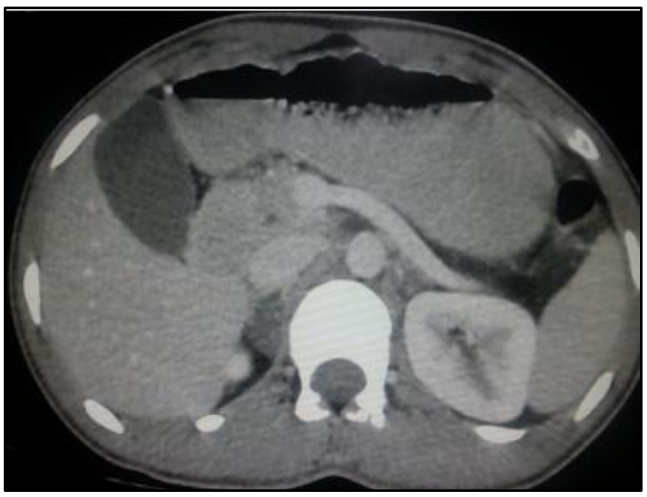

Figure 1

Figure 1. CECT axial section of abdomen venous phase reveals non-visualisation of pancreatic neck, body and tail and only globular shaped head of the pancreas is seen surrounding the 2nd part of the duodenum. The splenic vein is well visualised.

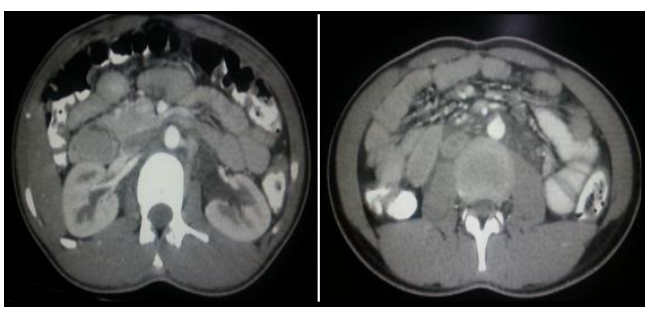

Figure 2 & 3

Figure 2 & 3. CECT axial section of abdomen at renal and infrarenal level showing soft tissue density mass lesion encasing the abdominal aorta and its branches extending into bilateral perinephric space (left > right). There is no sign of infiltration into abdominal aorta.

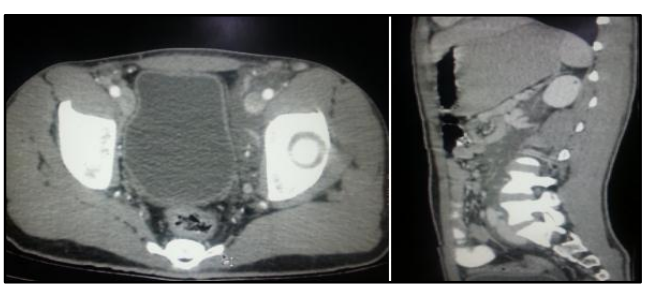

Figure 4 & 5

Figure 4 & 5. CECT axial and sagittal views of abdomen reveals the same soft tissue density mass lesion is seen encasing the iliac vessels on left side.

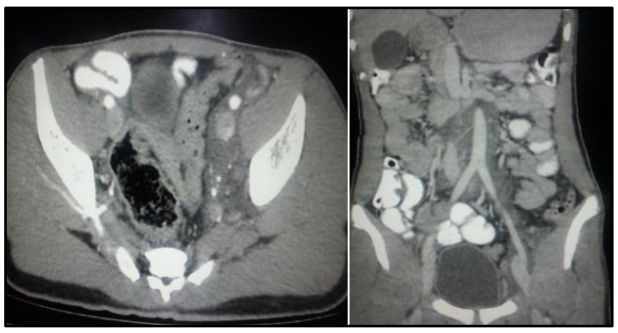

Figure 6 & 7

Figure 6 & 7. CECT axial and coronal reformat image showing the same soft tissue density lesion is seen encasing the abdominal aorta and inferior vena cava extending along the external iliac vessels on left side and is also seen extending up to the pelvis.

DISCUSSION

Abdominal filariasis presenting as an acute abdominal pain is very rare entity. USG and CECT scans of the abdomen demonstrated retroperitoneal lymphangiectasia. The CT imaging features of diffuse retroperitoneal lymphangiectasia have been described, but the confirmatory diagnosis can be made by the presence of microfilariae in the fluid aspirated from the cystic lesion. Though clinical and haematological examinations are thought to be inconclusive in a patient of acute abdomen, in our case diagnosis was solely made with the help of imaging modalities and demonstration of filarial parasites in ultrasound-guided aspirated cyst fluid from scrotal sac hydrocele.

Dorsal pancreatic agenesis is far more common than complete agenesis of pancreas, which is incompatible with life. Dorsal pancreatic agenesis is again subclassified into complete and partial pancreatic agenesis. In complete dorsal pancreatic agenesis entire neck, body and tail, the duct of Santorini (accessory pancreatic ducts), minor duodenal papilla are all absent, whereas in partial dorsal pancreatic agenesis a short rounded pancreatic head adjacent to the second part of the duodenum will be present, the size of the body of the pancreas varies, there will be remnant of the duct of Santorini and the major duodenal papilla will be present. Dorsal pancreatic agenesis is usually associated with other congenital variants such as polysplenia syndrome 1 which is most common, lobulation of liver, intestinal malrotation and heterotaxy syndrome 2. The clinical presentation of dorsal pancreatic agenesis ranges from complete absence of symptoms to recurrent pancreatitis, diabetes and sometimes exocrine pancreatic insufficiency. Diabetes is mainly associated with complete dorsal pancreatic agenesis and it is not associated with polysplenia syndrome.[1,4] Diabetes is seen in 67% of patient of complete dorsal pancreatic agenesis. Chronic and recurrent pancreatitis is seen mainly in partial dorsal pancreatic agenesis. It must be differentiated from pancreatic carcinoma, in which case there is upstream atrophy of the organ. Agenesis of the dorsal pancreas must be distinguished from a reduction in size of the pancreas due to autoimmune pancreatitis, chronic dorsal pancreatitis in pancreatic divisum and pancreatic atrophy secondary to focal pancreatitis of the body and tail of the pancreas. On MRCP if a dorsal duct of normal length is documented, then complete or partial agenesis of pancreas is ruled out.

CONCLUSION

The report highlights the importance of different imaging modalities in early diagnosis of retroperitoneal filariasis/lymphangiectasia with complete dorsal pancreatic agenesis. A timely ultrasound-guided aspiration of the cyst fluid and demonstration of the filarial parasite in it and demonstration of microfilaria in the peripheral blood smear confirmed the diagnosis. Retroperitoneal lymphangiectasia may present with acute abdominal pain. Because of early diagnosis, patient was placed on antifilarial drugs and has shown dramatic reduction of symptoms which denote earlier the diagnosis better is the prognosis of the patient. Complete dorsal pancreatic agenesis may be asymptomatic throughout life or it may present with focal pancreatitis and diabetes, although in our case report none of them were present and the patient was asymptomatic. By early diagnosing complete dorsal pancreatic agenesis at an early age patient will be aware of its condition and any future insult will be attended with caution and diligence.

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