TOTAL PANCREATIC LIPOMATOSIS – A RARE CAUSE OF MALABSORPTION

KEY WORDS: rv dysfunction, echocardiography, anterior wall myocardial infarction, inferior wall myocardial infarction.

Introduction
Pancreatic lipomatosis refers to replacement of pancreas with fat. It can be either focal or complete.

Focal fatty infiltration is more common than diffuse infiltration. The treatment for total pancreatic replacement s enzyme replacement with diet consultation; hence recognising the total pancreatic lipomatosis is very important in deciding the treatment plan for the patient.

Case report
A 35-year-old lady presented with complains of darkening of skin, weight loss and fatigue since 1 year. No history of decreased appetite. OGD was normal. Patient's symptoms have recurred and have progressive weight loss. There is history of intermittent steatorrhea on eating. No history of jaundice. No history of any bleeding. No history of TB, cough, asthma or hypertension. Investigations showed there is decrease in lipase levels.

Investigations
AMYLASE SERUM 121 U/L (norml value is < 200 U/L)
LIPASE 22 < 190

Literature review
Total pancreatic lipomatosis is a rare cause of malabsorption. The imaging modalities used for evaluating this rare entity is Ultrasound, CT and MRI.

Ultrasound can show increased echogenicity of the total pancreas. The disadvantages of the USG are it is subjective; bowel gas can obscure the visualization of the entire pancreas.

CT can reliably diagnose this condition. Non-contrast CT will show the pancreas as diffuse fat replacement of the pancreas.

MRI is more sensitive and can confirm the diagnosis. On T1 and T2 WI, the pancreas appears as diffusely increased signal intensity and it will be completely suppressed on STIR (fat suppression) sequence.

The pathogenesis of total pancreatic lipomatosis is not very clear. Common causes which predisposes to total pancreatic lipomatosis can be classified into local, systemic and syndromic associations.

Local causes
1. Chronic pancreatic duct obstruction by calculus in the pancreatic head[1]
3. pancreatic head adenocarcinoma after neoadjuvant chemo radiotherapy[3]

Systemic[4, 5]
1. Cystic fibrosis (most common cause in childhood)[4]
2. Atherosclerosis
3. Obesity
4. Steroid usage
5. Diabetes mellitus
6. Malnutrition
7. Hemochromatosis

Syndromic associations
1. Shwachman–Diamond syndrome (a rare autosomal recessive disorder characterized by exocrine pancreatic insufficiency, bone marrow dysfunction, leukemia predisposition, and skeletal abnormalities)
2. Johanson-Blizzard syndrome (abnormal development of the pancreas, nose and scalp, with mental retardation, hearing loss and growth failure)

In this patient, there is diffuse fat replacement of the pancreas noted with a tiny calculus noted in the pancreatic head within the main pancreatic head. In cases with main pancreatic duct obstruction due to calculus or tumor or stricture, complete fat replacement of the pancreas may occur. In animal experiments, ligation of the main pancreatic duct has been shown to produce degeneration and lysis of the pancreatic acini, with subsequent complete fat replacement of the pancreas[6].

Experimental and clinical studies have shown that fat replacement of the pancreas does result from ligation of the pancreatic duct or obstruction of the pancreatic duct by a tumor or a calculus. The pathophysiolog y of experimentally produced fat replacement of the pancreas has been described by Walters, using a minune model, where pancreatic ducts of rats were ligated.[7]

Conclusion:
Though total pancreatic lipomatosis is a rare cause of malabsorption, clinicians should rule out this entity if common causes are ruled out. CT can clearly depict the fat and can confirm the diagnosis. Pancreatic enzyme replacement with diet therapy can improve patient’s symptoms.

Figure 1
35-year-old lady with features of malabsorption; USG image shows increased echogenicity of the pancreatic head, neck, body and tail.

Figure 2
35-year-old lady with features of malabsorption; Computed tomography of the abdomen
a) Computed tomography of the abdomen obtained in venous phase through pancreas shows homogenous low attenuation suggestive of complete fat replacement of the entire pancreas.
b) Image obtained through the pancreatic head region shows a calculus in the main pancreatic duct; the distal part is not dilated.

Figure 3
35-year-old lady with features of malabsorption; Magnetic resonance imaging (MRI) of the abdomen

a) MRI – T1WI shows there is homogeneous increased signal intensity throughout the pancreas.
b) MRI – T2WI also shows there is homogeneous increased signal intensity throughout the pancreas.
c) MRI – T2WI with fat suppression shows there is complete suppression of the increased signal intensity of the pancreas – confirming the complete fat replacement of the pancreas.

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