Original Research Paper

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General Medicine

LIPOMATOUS PSUEDOHYPERTROPHY OF PANCREAS-AN EXTREMELY UNUSUAL CASE

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KEYWORDS :

INTRODUCTION

Lipomatous pseudohypertrophy (LPH) of the pancreas is an extremely rare condition in which there is an almost total replacement of native pancreatic tissue with lipomatous/adipose tissue. Peculiarly, there is no associated Obesity or diabetes or symptoms/signs of pancreatic inflammation. We describe such a case from our institution.

CASE REPORT

Mrs PP was a F/68 years female with no significant past illnesses (besides COVID 3-4 weeks ago from which she had made an uneventful recovery) and on no regular medications. She presented with a pain in epigastrium which had begun from 2-3 months. Initially mild, it had built up lately to a "moderate to severe" intensity. It invariably started after food and fearing this, the patient had started avoiding food. There was also constant nausea with a strong underlying anorexia. She invariably vomited after eating and had lost 5-6 kgs in 2-3 months. Besides normocytic normochromic anaemia, her previous investigations showed no other abnormalities. Her OGD's copy had been done few weeks ago and was declared positive for H.Pylorii. She had been given Triple drug therapy, with no apparent benefit.

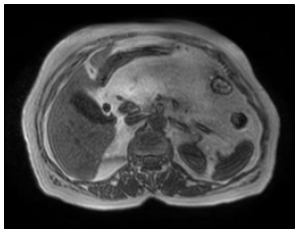
O/E She was comfortable in bed, BP-120/80, P-80/m, SpO2-99% on room air and appeared pale. There was no icterus/ Clubbing/ Lymphadenopathy. Abdomen was soft with no organomegaly. There was mild to moderate tenderness on deep epigastric palpation. Following is the table of current investigations.

Table of investigations

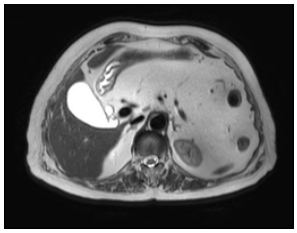
Hb	11.4
WBC	10,700
Platelets	279
ESR	21
CREATININE	0.68
SGPT SGOT	26 32
AMYLASE	40
HbA1C	6.2
TSH	2.11
Stools	Normal
Stools-fat globules	Not detected
CA-19.9	20.28 (WNL)

Radiology: findings and D/D

MRI showed marked enlargement of pancreas and diffuse replacement of its parenchyma by fat. This was established by T1 and T2 hyper intensity identical to the mesenteric / subcutaneous fat and loss of signal on fat suppressed sequence. The traversing vessels within were not distorted. No soft tissue or abnormal enhancement was identified. There was mass effect on the gastric antrum and the C-loop of duodenum.



1 a.



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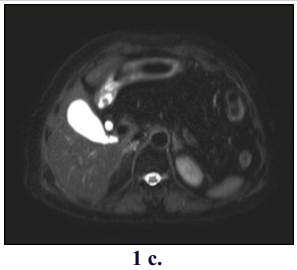

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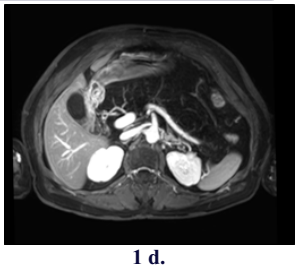

1 d.

Fig1. a – T1, b – T2, c- T2 fat suppressed, d – T1 Post contrast MIP

These features are typical for lipomatous pseudo hypertrophy of pancreas[1].

In view of the classical radiological features histopathological confirmation was not deemed necessary[1].

Simple fatty infiltration of pancreas is much more commonly encountered. But in LPH this fatty replacement is to such an extent that there is pancreatic enlargement and mass effect.

Retroperitoneal liposarcomas are known to contain fatty tissue, but they show nodular soft tissue or enhancement. Vessel tissue distortion would also be expected. None of these features were evident in this case.

Diagnosis

Lipomatous pseudo hypertrophy of Pancreas.

DISCUSSION

Aetio pathogenesis

Lipomatous pseudo hypertrophy of pancreas is an extremely rare entity. Less than fifty (authentic and confirmed) cases have been reported worldwide.[2]

In this situation, there is a replacement of the entire pancreas with huge amounts of fat/adipose tissue with consequent uniform enlargement of the pancreas. This "enlargement" is not a real bona fide hypertrophy of the actual pancreatic contents but that of the fatty tissue within. Hence the nomenclature of "Lipomatous pseudo hypertrophy". However, the shape of the pancreas is usually preserved. The pancreatic exocrine tissue is hugely depleted and is replaced by adipose tissue. Peculiarly the ducts and islets are largely preserved.[3]

This condition was first described by Hantelmann in 1931.[4]

Usually, the entire pancreas is affected (in as many as 70% of cases). Rarely, there may be localised involvement of the Head/body/ tail of pancreas.

It might be congenital or acquired. However, the exact aetiology and pathogenesis both remain elusive. It has been hypothesised that a viral aetiology is likely. The extremely low number of reported cases makes it difficult to prove or disprove this.[5]

There has been a noted association with association with disorders like cystic fibrosis,Shwachman-Bodian-Diamond syndrome,Bannayan syndrome and Johanson-Blizzard syndrome. This entity must be

differentiated from the adipose tissue infiltration of pancreas that is seen in conditions like morbid obesity, Type 2 diabetes mellitus, or chronic pancreatitis.[6]

Initially most of the cases were described in younger population (adolescents/ children). However, lately, older/middle aged persons too are being reported.[7]

There was a renewed interest in "inducing" lipomatous pseudohypertrohy of pancreas in experimental animals. Toxic agents such as ethionine or certain viruses, when introduced in pancreas of experimental animals, were successful in producing a diffuse and massive fat replacement, which largely mimicked the LPH of the pancreas in humans. Chronic hepatitis B too has been suspected as a possible agent.[8]

Clinical features

It may be asymptomatic in a huge majority of patients, being discovered incidentally during routine evaluation. When it does manifest clinically, the usual features are

(1) A dull/ boring abdominal pain that usually appears post prandially.
(2) In case there are features of exocrine insufficiency, there may be bloating, flatulence, Diarrhoea with foul smelling stools and abdominal heaviness/discomfort.
(3) If there is a massive hypertrophy, there may be compression of Bile duct, causing obstructive jaundice.
(4) There have been instances of attending Doctors mistaking this mass as a pancreatic carcinoma.

Course of our patient during hospital stay

She was reassured and told to have only liquids initially. IV DNS (40 ml/hr) was started with multivitamin and D3 supplementation. Later, after evaluation was complete, Rabeprazole 20 mgs OD and pancreatic enzyme (Pancreatic Minimicrospheres Capsules-10,000) was added.

Once her diagnosis was established and she was convinced that there was no underlying malignancy and no Surgery/ chemotherapy was being considered, she relaxed to a huge extent. We increased her oral intake and encouraged escalation to semi solid food. On tolerating this, she was instructed to start normal (sugar restricted) diet, which she could successfully manage. IV fluids were weaned off. She was discharged on day 5 of admission.

OPD Follow up

On reassessment after 3 weeks, she reported resolution of her symptoms to a huge extent. She had started eating normally and her weight had gone up by 2 kgs. She continues to be on pancreatic enzyme supplementation and her sugars are being monitored on regular basis.

CONCLUSION

We describe here an extremely rare entity that presents a tremendous diagnostic challenge. Despite a wide range of clinical symptoms, there is a marked paucity of haematological and biochemical disarrays. Though USG of abdomen might point towards the correct diagnosis, it's only an MRI that's conclusive. Treatment too is mostly supportive/ symptomatic, as overly aggressive treatment would be counterproductive. The talent would lie in correct diagnosis and intervention only when absolutely needed, as surgical traumas might do more harm than good.

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