SITUS INVERSUS TOTALIS WITH PANCREATIC TAIL ADENOCARCINOMA – A RARE CASE REPORT

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ABSTRACT:-Situs inversus is a characterized by the transposition of organs to the opposite side of the body. When associated with dextrocardia, it is known as situs inversus totalis. This condition is rare congenital abnormality and can create diagnostic confusion for surgeon due to unusual location of symptoms leading to late diagnosis of known pathologies as well as operative difficulties due to altered anatomy. Pancreatic tail malignancies are notorious for their nonspecific signs & symptoms leading to late diagnosis and poor prognosis. We report a rare case where these conditions coexisted. A seventy year old man presented with right sided abdominal pain since three months& vomiting since one month. X-ray chest showed dextrocardia. Ultrasonography& Computerised Tomography (CT) scan were suggestive of solid cystic lesion in tail of pancreas with situs inversus totalis. Distal pancreatectomy with splenectomy was performed. Patient made uneventful recovery and is doing well at one year follow up.

KEYWORDS:-Situs inversus totalis, Adenocarcinoma pancreas, Distal pancreatectomy with splenectomy

INTRODUCTION:-Situs inversus is a rare congenital condition in which the major visceral organs are reversed or mirrored from their normal positions. The reversal of the organs may then lead to diagnostic confusion [1]. Mass lesions in body & tail of pancreas though less common, are ominous due to their late presentations [2]. This can be further delayed in patients with Situs Inversus due to unusual presentation hence this case report.

CASE REPORT:- A 70 year old man presented with complaints of right sided abdominal pain which was dull aching, not related to meals since three months &vomiting since one month. There was no loss of weight or appetite. Pain was not associated with jaundice. On examination patients vitals were stable and there was no icterus. Abdomen was soft with no palpable lump. X-ray abdomen was normal. Ultrasound abdomen was suggestive of well defined thick walled cystic lesion from tail of pancreas with central necrosis. Roentgenogram of chest diagnosed presence of situs inversus. Computerized tomogram (CT) of abdomen showed 10x5x6 cm solid cystic neoplasm in tail of pancreas with Situs inversus totalis. Serum CEA & Serum 19-9 were elevated. Endoscopic ultrasound

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was suggestive of cystic lesion in tail of pancreas 10x7 cm with eccentrically thickened walls & clear content separate from spleen. Patient was subjected to a midline laparotomy. Distal pancreatectomy with splenectomy was performed in view of dense adhesions between mass and spleen. Patient made uneventful postoperative recovery. Final histopathology was suggestive of poorly differentiated adenocarcinoma of pancreas with free resection margins. Patient was discharged on postoperative day 10 and is well at 1 year follow up.

DISCUSSION:-Situs inversus (also called Situs transversus or oppositus) is a condition in which the organs of the chest and abdomen are arranged in a perfect mirror image reversal of the normal positioning. It may or may not be associated with dextrocardia, also known assitus inversus totalis. First known case of situs inversus in humans was reported by Fabricius in 1600. The incidence is thought to be 1:5000 to 1:20000.Its transmission mode is autosomal recessive, but its precise genetic mechanism has yet to be identified. It is associated with a number of other conditions such as Kartagener's (bronchiectasis, sinusitis, situs inversus) and cardiac anomalies.

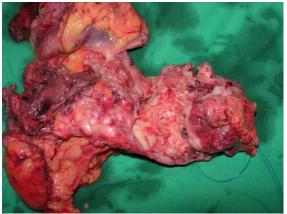
About 25% of individuals with Situs inversus have an underlying condition known as primary ciliary dyskinesia (PCD).Dysfunction of cilia causes increased susceptibility to lung infections. Although only 3-5% of people with Situs inversus have any type of functional heart defect, this is higher than the rate of heart defects in the general population, which is less than 1%.Situs inversus has been found to be associated with dysplasias & cysts in pancreas and kidney ^[4]. The prognosis for an individual with Situs inversus is good, and in the absence of a heart defect or other underlying diagnosis, life expectancy is normal. There is no treatment for Situs inversus. However, because of altered anatomy patients present with unusual symptoms of common pathologies. This might lead to diagnostic confusion and delay in diagnosis. Also it may lead to difficulty during surgery. Situs inversus also complicates organ transplantation operations as donor organs will almost certainly come from Situs solitus (normal) donors.

Patients with tumors arising in body or tail of pancreas usually do not develop jaundice or gastric outlet obstruction ^[5].Weight loss & abdominal pain are usual presenting features. Thus they are diagnosed late & have poorer prognosis ^[2]. Situs inversus might just further add to diagnostic problems. Only 10% of cancers involving body & tail of pancreas are resectable at diagnosis. 5 year survival in patients who are resectable is 8-14%. ^[5]. Pancreatic cancer serum markers i.e. Sr. CEA & Sr. C.A. 19-9 might not be raised in early curable form of disease ^[5]. Transcutaneous ultrasound followed by contrast enhanced C.T Scan is most commonly used imaging modalities. ERCP & EUS might be helpful ^[5]. Distal pancreatectomy with or without splenectomy (recommended for malignant tumours) is treatment of choice in resectable tumours.

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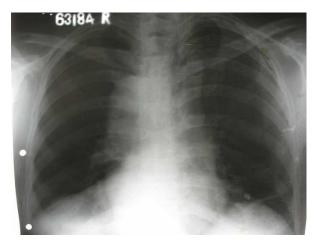
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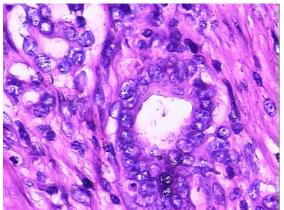
Intra-op photograph of the resected specimen



Pre-operative CT image showing situs inversus totalis



Chest x-ray showing dextrocardia in the same patient



Histo-pathology of pancreatic adeno-carcinoma