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AN INCIDENTAL DETECTION OF SITUS AMBIGUOUS WITH LEVOCARDIA (HETEROTAXY SYNDROME) WITH ABDOMINAL MANIFESTATIONS: A RARE CASE REPORT

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ABSTRACT

While dealing with a case of Situs Ambiguous abnormality, there is a need of proper terminology in making a specific diagnosis and describing associated anomalies .Moreover there is need of understanding the detailed study about heterotaxy syndrome, the left and right isomerism and differentiation between asplenia and polysplenia. Our case is presented as a case of Situs Ambiguous with Levocardia in an adult Female (aged 47 years) associated with abdominal manifestations like midline liver with Gall Bladder in left para median location with stomach and multilobed spleen,descending colon on right side of Abdomen and caecum , I/C junction on left side of abdomen, suggestive of Intestinal Rotation.Overall our case reveals many unique findings on ultrasound and C.T.Scan Abdomen associated with Situs ambiguous anomaly which allows for further categorization and characterization of situs abnormalities and their significance.

KEYWORDS

Situs Ambiguous, Levocardia, Polysplenia, Heterotaxy

AIM:
It is a Retrospective study of a rare case report of situs Ambiguous with Levocardia in an adult female.

Our aim is to describe the role of imaging modalities like ultrasound, C.T. Scan Abdomen in detection of accurate diagnosis and describing associated anomalies.

OBJECTIVE:
The main object of our study was early and accurate diagnosis of such rare congenital anomalies detected incidentally when patient presented with other disease on the basis of sign, symptoms and imaging modality findings and at the time of surgery planning so as to prevent any complications

INTRODUCTION /HISTORY (BACKGROUND):
Though adequate literature is available on imaging features of situs abnormalities

In pediatric population because most of children present with these abnormalities with severe congenital heart disease, immune deficiency and bowel obstruction as a result of malrotation.

In contrast, there is a scanty information available in radiology literature about incidental detection of abnormal abdominal manifestations without cardio-pulmonary and vascular defects in situs abnormalities in adult.

Infact, Situs abnormalities are usually diagnosed incidentally in adults during imaging evaluation in patients with abnormal gastro-intestinal and cardiac disorder and any unrelated medical conditions.

Now a days, recent advances in diagnostic imaging technique like high resolution sonography ,C.T.scan ,Magnetic Resonance Imaging and Angiography have greatly increased our efficiency in diagnosing these situs abnormalities.

The subcategory of situs Ambiguous with polysplenia which is asscociated with less severe or no congenital heart disease would be detected incidentally more often.

In this article, we will discuss about the abdominal manifestations of situs anomalies in adults.

CASE REPORT:
The Patient presented initially with nausea and pain in left hypochondrium and consulted Local Physician at Hapur,.The ultrasound abdomen was advised .On ultrasound report , radiologist reported cholelithiasis with liver and gall bladder deviated to left side . Ultimately she came to attend surgical OPD at G.S.Medical College & Hospitals and advised ultrasound again.On Ultrasound examination , there was calculi in gall bladder located towards left side of midline and liver was found on left hypochondrium side while spleen was situated on right hypochondrium and caecum on left iliac region.

C.T Scan Abdomen was advised by surgeon prior to planning surgery and for confirmation of ultrasound findings.

IMAGING FINDINGS:
The X-Ray Chest of patient showed cardiac Apex and Aortic Knuckle on left side which was further confirmed by the findings of c t scan where cardiac apex seen on left side chest, suggestive of Levocardia .Both the lungs were found normal. Cardiac size was found of normal size so ruled out any possibility of Cardio-pulmonary anomaly .The gas shadow found in right hypochondrium, creates suspicion of stomach on right side.

Due to having suspicious findings of situs abnormalities on X ray and ultrasound, patient underwent C T Scan abdomen for exact characterization of situs abnormality.

The C.T.Scan Abdomen revealed many abnormal findings such as liver located in midline with a notch on inferior surface of right lobe of liver and left lobe of liver seen in left hypochondrium .Gall Bladder was situated in left paramedian location .cardiac apex is seen on left side suggestive of Levocardia ,further confirmation was done by echocardiography also.

The contrast filled stomach was seen on right side with indenting right lobe of liver and bilobed spleen noted on infero-lateral aspect of stomach on right side .

The Caecum is demonstrated in left iliac region while descending colon is demonstrated In right iliac region ,suggestive malrotation of gut.

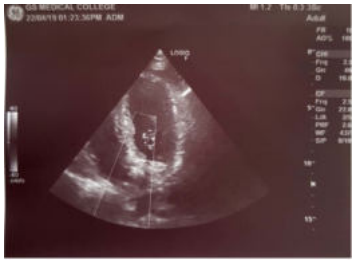

Image 1: Ultrasound reveals calculi in gall bladder lumen, located left paramedian region . Liver is seen in left hypochondrium .

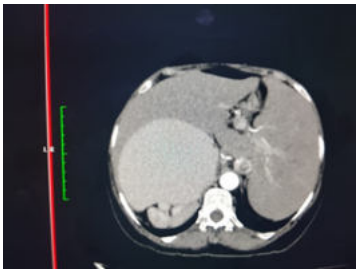

Image 2:C.T.Scan Abdomen demonstrate liver on left hypochondrium and stomach in right Hypochondrium with bilobed spleen below stomach.

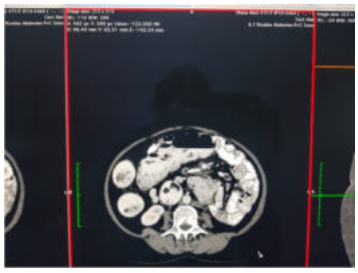

Image 3: this slice of C.T.Scan abdomen reveals caecum in left iliac region and ascending colon on left side .

INTRA-OPERATIVE FINDINGS:
After getting confirmation of imaging findings and deep discussion between radiologist and Surgeons, they decided to operate the gall bladder calculi by laparoscopic Cholecystectomy, during surgery, surgeon found Right lobe of liver located in left hypochondrium while left lobe is crossing the midline and reaching in right side of abdomen and gall bladder in left paramedian position.

The Stomach was demonstrated on right side with bilobed spleen detected on right hypochondrium .Caecum and ascending colon is seen in left iliac region.

Uterus was not seen as there was past history of hysterectomy.

FOLLOW-UP:
Post-operative follow up ultrasound reveals non-visualization of Gall bladder and uterus .No evidence of any distension /or fluid collection is seen in abdominal cavity .Patient was found normal and devoid of any complaint so discharged and advised to attend surgical OPD in case of any symptoms as and when required.

TERMINOLOGY:
Due to over lapping features of various types of situs anomalies ,the term situs is used in relation to position of heart and viscera relatives to the midline .

Various situs anomalies have been described like situs solitus, situs inversus and situs ambiguous.

When situs cannot be determined between situs solitus and Situs Inversus, the patient is categorized into 3rd category i.e. Situs Ambiguous or heterotaxy syndrome.

In these patients, the liver is located in midline, may be right or left side dominant, the spleen may be Single or bilobed, multiple or may be absent and the bowel may be normal or malrotated.

The two main sub type of situs ambiguous include:-
1. Right Isomerism or asplenia syndrome.
2. Left Isomerism or polysplenia syndrome.

In classic right isomerism or asplenia or lvermark syndrome – bilateral right sidedness occurs. The patients have bilateral right atria, a centrally located liver, and an absent spleen. Both the lungs have three lobes, the descending aorta and inferior vena cava are on the same side of the spine.

Right isomerism has an incidence of between 1 in 10000 and 1 in 20000 births, with male predominance.

And nearly 100% incidence of CHD and often present in childhood with cyanotic Heart disease.

While in left isomerism, there is bilateral left sidedness. These patients may have bilateral left atria and multiple spleens. Both the lungs have 2 lobs. Interruption of the vena cava with azygous and hemiazygous continuation is often present. The features of situs ambiguous cases are challenging and require through evaluation of the viscera. (1)

DISCUSSION:
Heterotaxy syndrome is a rare, complex and more confusing type of the situs anomalies .It is very difficult to estimate the degree of lateralization, isomerism and rotational variation in both types of cases.(2)

There is no single type of specific condition or particular pathognomic findings for left sided isomerism with polysplenia. Abdominal organs like liver is midline or right or left dominant,Gall bladder may be left paramedian,there may or may not be gastro-intestinal rotational anomalies. Spleen is on the right side and may be multilobed.

The majority of patient's up to 90% have less number of congenital heart diseases.

Asymmetry or unilateral organs develop from the embryologic midline structures

Although situs ambiguous cases are rarely seen, various anomalies in varying degrees related to intra- abdominal, cardio-pulmonary and vascular structures are observed in these cases

Solid intra-abdominal organs may be located in midline

Applegate et al (3) already reported a series of the ten polysplenic patitients for heterotaxy syndrome.

To our best knowledge, the case we present does not correlate one to one with any of the cases reported earlier, so unique in itself, situs ambiguous with levocardia with abdominal manifestations only without any cardio-pulmonary defect detected incidentally in an adult female of 47 years age, consulted physician for pain in abdomen.

By using imaging modalities like echocardiography, ultrasound, C.T.Scan, M.R.I. and Angiography, it is now possible to reveal these variations prior to surgery to prevent the possible risk and complications. (4)

We suggest that the term heterotaxy syndrome be used to describe these patients, rather than using imprecise terms like asplenia or polysplenia.

The heterotaxy syndrome is much more descriptive than polysplenia.

A more accurate classification in patients with Heterotaxy is desirable in those adult patients with prolonged survival who are at increased risk of congenital Heart disease, immune deficiency and intestinal rotational anomalies. (5)

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Conflict of interest --Author declare no conflict of interest.

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