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"A Case Report"- 'A Rare Case of CML in a Patient with Situs Inversus Totalis'.

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Abstract:
A 50 years of age female presented to the General Medicine opd with complaints of easy fatiguability, abdominal distension, fever (on and off) and weight loss for past 3 months. Admitted and evaluated her in detail. Her Chest X-ray showed f/s/o Dextrocardia. Usg abdomen & pelvis showed situs inversus totalis. Due to elevated white blood cell count , as well as platelet count and decreased Hemoglobin, patient was planned for Bone marrow aspiration and biopsy. Both confirmed presence of CML in accelerated phase. P210 positive for BCR-ABL transcript. Imatinib mesylate administered. Following which her white blood cell count started decreasing. Patient showed improvement in her general condition. Patient is under regular follow up . She is on Imatinib mesylate 400 mg 1 tablet daily. Situs inversus totalis is a congenital anomaly, in most cases remains asymptomatic and occurs in concomitant with other disorders. There are very few reports depicting association between cancer and situs inversus totalis. Our case study is one among them, which shows some association between CML and Situs inversus totalis.

Key Words: *CML, Situs inversus totalis, P210 positive* **.**

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I. Introduction:

Situs solitus is the normal situation of major visceral organs. Situs inversus totalis which is also called as situs transversus/oppositus is a congenital anomaly(Autosomal recessive) due to failure of embryonic cells to rotate causing thoracic and abdominal visceral organs being mirrored from their normal position. Mostly the persons affected by this anomaly remains asymptomatic. 1 person in 10,000 seems to be affected . Isolated levocardia uncommon. There is 5 to 10 % prevalence of heart disease- Transposition of great vessels. Most probably its an incidental diagnosis. Associated with syndromes like Kartagener's syndrome etc. Diagnosis is made using Chest x-ray, USG abdomen and pelvis, CT/MRI.

Chronic myeloid leukemia is a clonal haematopoietic , myeloproliferative stem cell neoplasm. Philadelphia chromosome (reciprocal balance translocation between long arms of chromosomes 9 and 22 {t 9,22} -BCR ABL 1) is seen in > 90% of patients with CML. Course of CML divides into – Chronic phase, accelerated phase and blast phase. Male predominance seen. Median age group affected- 55 to 65. Worlwide annual incidence is 2,00,000. CML is considered to be 15% among all leukemias. Ionising radiation is the most common etiology. Clinical features of CML are- fever, fatigue, symptoms of anemia, symptoms of thrombosis, abdominal pain/distension, weight loss etc. Signs of CML: Anemia, splenomegaly, hepatomegaly, lymphadenopathy. Sokal score categorises CML into- low(60-65), intermediate(25-30) and high(10).

Diagnosis of CML made using FISH and PCR which depicts the presence of BCR-ABL 1 fusion gene. 10 year survival is 85% with First generation-Imatinib mesylate introduction. Second generation drug- Nilotinib and Third generation- Ponatinib can also be used as alternatives. Nilotinib causes QTc prolongation as side-effect. Allogenic stem cell transplantation is also beneficial.

Very few cases have been reported of this sort, association of CML with Situs inversus totalis.

II. Case Report:

A 50 years of age female presented to our opd with complaints of easy fatiguability, fever, abdominal distension and weight loss for past 3 months.On examination: Her vitals stable. Pallor+. Massive splenomegaly+. She was admitted and subjected to various investigations. Her chest x ray and ECG showed presence of right sided heart. Usg abdomen and pelvis showed – situs inversus totalis. Complete blood count

showed- Leukocytosis, thrombocytosis and anemia. Bone marrow aspiration and biopsy done, which supported the diagnosis of CML in accelerated phase(myeloid>erythroid).FISH and PCR assays done which identified the BCR-ABL 1 FUSION GENE. Imatinib mesylate 400 mg/day initiated. WBC started decreasing and her general condition seemed to improve. Patient is on Imatinib and under regular follow up.

META-DATA:

CBC:HB- 6.8, WBC- 125000, PLT- 428000, HCT-22.2, RBC- 2.36 MILLION, MCV-93.3, mch-28.6, mchc-30.6, g-68, l-12, m-19, b-1, esr-120

CHEST X RAY & ECG: Right sided heart features

USG ABD&PELVIS:F/S/O SITUS INVERSUS TOTALIS

P.S.: RBC- Normocytic, microcytic hypochromic anemia. WBC- N- 55, Metamyelocytes- 10, blast- 10, e- 2, b-3, PLT- adequate.

BCR-ABL STUDIES

(FISH + PCR): P210 positive for BCR-ABL transcript. t(9,22) found.

BONE MARROWASPIRATION & BIOPSY suggests: CML IN ACCELERATED PHASE.

RBS; 143

LFT: S. Bilirubin- 0.7, s. ALP- 272, S. albumin- 3.2, total prot- 7.9, sgot- 67, sgpt- 30.

S. URIC ACID- 7.4

S. LDH: 210

S. MG2+: 1.9

RFT: U- 33, C- 0.5

S. ELECTROLYTES: Na+: 135, K+: 5.2, Calcium- 9.4, S. PO42-: 4.4, S. Cl-: 95.

BLOOD GROUP: B positive.

III. Discussion:

Situs inversus totalis is not a premalignant condition. 4 most common cancers associated with situs inversus totalis are- gastric cancer, colorectal cancer, lung and kidney cancers. Certain studies have showed possible association between Situs inversus totalis and cancers. Previous studies shows kinesin – to play an important role in situs inversus totalis pathology. Chronic myeloid leukemia abbreviated as CML is a cancer of white blood cells. It is a form of leukemia characterized by the increased and unregulated growth of predominantly myeloid cells in the bone marrow and the accumulation of these cells in the blood. It is a clonal bone marrow cell disorder in which there is a proliferation of mature granulocytes (neutrophils, eosinophil, and basophils) and their precursor associated with a characteristic chromosomal translocation called the Philadelphia chromosome. Our study explains the possible link between CML and situs inversus totalis.

IV. Conclusion:

The present study concluded that association of CML with situs inversus totalis may be by chance. Our report emphasizes on the fact of the need of good clinical evaluation by a qualified therapist and use of appropriate investigative studies is mandatory in order to secure patient from such a critical health condition

ETHICS COMMITTEE: Since the study is a case report, Ethics committee approval not required.

CONSENT: Patient's informed consent obtained for publishing her details.

Funding: Nil

DISCLOSURE: The Author has no conflicts of interest.

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