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RESEARCH ARTICLE

A CASE OF HODGKIN LYMPHOMA PRESENTING AS NEPHROTIC SYNDROME

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Abstract

Paraneoplastic glomerulopathy has been reported in patients with malignancy.In particular, the association of Nephrotic Syndrome (NS) with Hodgkin's Lymphoma (HL) is rare and there are few reports in the literature. Minimal change nephropathy is the most frequently observed renal lesion whereas this association appears, either simultaneously or within several months of each other. Nephrotic changes as part of the paraneoplastic syndrome are rare in lymphoid malignancies. 1% of cases of Hodgkin's Lymphoma cases may present with nephrotic syndrome. We are reporting a case which initially presented as nephrotic syndrome and later was diagnosed to have of Hodgkin's Lymphoma and managed accordingly. This report presents the case of a 13-year-old male in whom the diagnosis of NS was established six months before the diagnosis of Hodgkin lymphoma. Once chemotherapy was initiated, proteinuria and the clinical manifestations of NS, mainly oedema, disappeared. Considering that the NS can be a paraneoplastic manifestation of Hodgkin lymphoma, the careful clinical evaluation becomes mandatory in any child with NS and persistent proteinuria despite appropriate treatment protocol. The prognosis of these patients is good, the treatment of Hodgkin disease causing the disappearance of proteinuria.

Introduction:-

Nephrotic changes as part of the paraneoplastic syndrome are rare in lymphoid malignancies.In particular, the association of Nephrotic Syndrome (NS) with Hodgkin's Lymphoma (HL) is rare. Minimal change nephropathy is the most frequently observed renal lesion whereas this association appears, either simultaneously or within several months of each other. Since 1922 the association between NS and extra renal neoplasia was described by Galloway et al. (1).To date, the literature addressing this relationship consists of only a limited number of case reports and case series in which NS, as paraneoplastic syndrome, is linked to different malignancies such as leukaemia, Hodgkin lymphoma, non-Hodgkin lymphoma and different carcinomas9(2). Particularly, it was observed the association between minimal change nephropathy and Hodgkin's lymphoma, these two entities emerging either simultaneously or within several months one from the other (3-6). The accurate basis of this relationship rests unknown, even though there have been hypotheses regarding a T-cell dysfunction (7).

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Case Report

A 13-year-old, boy was admitted with complaints of Abdominal distension, periorbital and pedal edema. On Evaluation, patient had Normal Renal function tests with proteinuria and hypoalbuminemia with ultrasound abdomen showing normal kidney size and an initial diagnosis of Nephrotic syndrome was made and started on prednisone. He achieved remission in 2 weeks. After 6 weeks of daily steroids he was put on alternate day prednisolone. 4 months later his symptoms reoccurred and on ultrasound abdomen he had multiple enlarged and matted mesentric lymphnodes. He was started on oral steroids and in view of matted lymphadenopathy, empirical anti tubercular therapy was started and discharged after 5 days as he was in remission and advised to follow up. After 2 months, his symptoms reoccurred, he also developed fever and cervical lymphadenopathy. General examination showed pallor and edema. vitals :PR- 126/min, Blood pressure- 100/60 mmHg , RR- 18/min , Temperature -101^0 F. On Respiratory system examination, decreased breath sounds heard. GIT examination revealed abdomen distended , no organomegaly and no tenderness. Cardiovascular and nervous system examination were normal.

Investigations

CBP Hb - 5.2gm%, TC-9400 cells/mm^3, Platelet count- 3.26lakhs/mm^3, Differential count - P89 L8 E2 M1, **ESR** - 70mm/1st hr,

Liver function tests:

Total Bilirubin - 1.3mg/dl (Direct-0.5, Indirect-0.8) SGOT-39 IU/lit SGPT-8 IU/lit ALP-59 IU/lit, Serum Protein - 4.1g/dl (A-2.4,G-1.7)

Renal function tests:

Serum creatinine - 0.3mg/dl, Blood urea - 33mg/dl, 24 hr urine protein -28mg/2000ml

Serum electrlytes:

Na-137meq/lit,k- 3.46meq/lit, Cl-99mew/lit.

QBC –

negative

USG Abd –

B/l pleural effusion , minimal ascites, B/l mild HUN , Retroperitoneal and mesentric lymphadenopathy

CECT Chest and Abdomen –

E/o multiple oval to round soft tissue density lesions -S/o Lymphadenopathy

USG Neck –

B/l cervical lymphadenopathy

Exicision biopsy from cervical LN –

scattered large cells with vesicular nuclei, prominent nucleoli and moderate amounts of cytoplasm resembling mononuclear Hodgkin cells and Reed-Sternberg cells S/o Classic Hodgkin lymphoma, mixed cellularity type

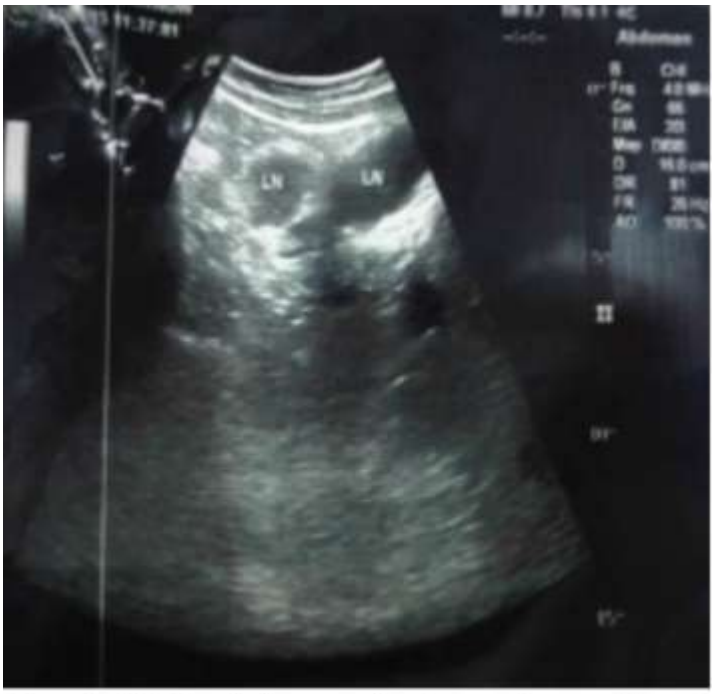

Fig 1: USG abdomen showing retro peritoneal lymphadenopathy

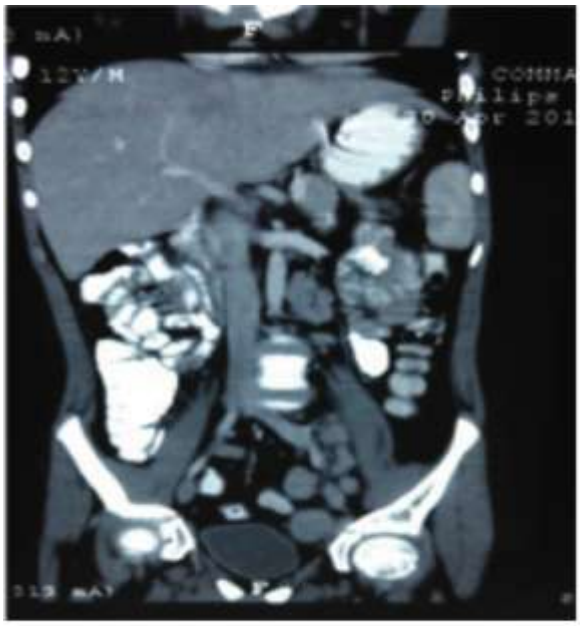

Fig 2: CECT abdomen showing para aortic & iliac lymphadenopathy

Discussion:-

The relationship between NS and Hodgkin lymphoma is well known. Consequently, attention must be paid to the presence of proteinuria as a sole element of a paraneoplastic syndrome. The careful examination, in order to exclude malignancies, becomes mandatory when facing a patient with NS. Chemotherapy once initiated, will determine the remission of NS, the collaboration between the pediatric nephrologists and oncologists being of utmost importance (8).

The exact incidence of NS in patients with Hodgkin lymphoma remains unknown. Incidence of 1% and 0.6% respectively was reported in two large series from France and Turkey – 9 out of 1144 children with Hodgkin lymphoma in both studies (6,9). Between 1984 and 2015, 136 patients with Hodgkin lymphoma were treated in our clinic, the case presented here being the second in which these two pathologies co-exist. To the best of our knowledge, these two cases (1.6%) are the only ones presented in Romanian paediatric population, the first case being published by Scurtu C. et al in 1998 (10).

Stephan et al. analysed the prevalence of NS in patients diagnosed with Hodgkin's lymphoma and found that 5 out of 483 children suffering from Hodgkin lymphoma, followed for a period of 13 years, developed nephrotic range proteinuria. This group of research concluded on the basis of their case series that thorough evaluation of patients with Hodgkin lymphoma is mandatory because, even though rare, glomerular dysfunction can be present (6).

In 2010, Farruggia et al. published two case reports where they analysed patients who had the association Hodgkin lymphoma - nodular sclerosis and NS - minimal changes nephropathy. Both patients received chemotherapy which consisted in the administration of 6 COPP/ABV cures plus radiotherapy, observing that 4 years after the end of the treatment patients maintained remission of both diseases (11). These results are in accordance with the literature data where in patients with this unusual association, the most prevalent subtype of Hodgkin's lymphoma, in both adults and children, was nodular sclerosis (6,9) and the predominant form of nephrotic syndrome was the minimal change nephropathy (4,5). Nonetheless, it is to be mentioned that 0.4% of patients with Hodgkin lymphoma present this histological feature of NS (12).

Geeta Gathwala et al. published in 1994 a case similar to ours, in which a 6 year-old boy was admitted in the hospital for right laterocervical tumefaction evolving for about 9 months, and generalised oedema developed progressively over the past 10 days. Laboratory data revealed nephrotic range proteinuria and severe hypoalbuminemia, sustaining the diagnosis of nephrotic syndrome. Biopsies of the lymph node concluded on Hodgkin lymphoma – nodular sclerosis and chemotherapy was initiated with very good response and remission of proteinuria after the first two cures (13).

In 2002, Raphael et al. published two case reports with Hodgkin's lymphoma and secondary NS, underlining the fact that chemotherapy determined the remission of the NS. They also observed that recrudescence of proteinuria was associated to Hodgkin's lymphoma relapse (14).

Nephrotic syndrome can develop before or after the diagnosis of Hodgkin lymphoma. In one of his studies, Audard et al. found that 38% of patients developed NS before the diagnosis of lymphoma, 43% after, and 19% develo ped the two diseases simultaneously (15). Also, Stephan et al. presented two patients in whom NS preceded with 6 months and 12 months respectively the diagnosis of lymphoma and o ther 3 patients where the two diagnosis where made at the same time (6).. To date, the longest interval reported between these two entities was 42 months (4).

Conclusion:-

Considering that NS can be a paraneoplastic manifestation of Hodgkin lymphoma, careful clinical evaluation becomes mandatory in any child with NS and persistent proteinuria despite appropriate treatment protocol. The prognosis of these patients is good, the treatment of Hodgkin's disease leads to the disappearance of proteinuria.s

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