LANGERHANS CELL HISTIOCYTOSIS OF SMALL INTESTINE

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PRESENTATION OF CASE

A 55-years old female patient with no known comorbidities presented with chief complaints of pain abdomen for 3-months associated with vomiting, abdominal lump and backache for past one month. Pain was insidious in onset, moderate intensity, originated in periumbilical and epigastric region and was relieved to some extent by oral analgesics. Abdominal lump developed in periumbilical region gradually increased in size upto the size of a tennis ball & was associated with abdominal distention and non-passage of stools & flatus. She was a chronic beedi smoker for 30-years. Contrast enhanced computerized tomography revealed an ill-defined growth in abdomen causing intestinal obstruction. She eventually got operated for sub-acute intestinal obstruction. She was a chronic beedi smoker for 30-years. On general physical examination patient was cachexic, ECOG status-3 and was taking liquid diet only. Local examination revealed tense and distended abdomen with unhealthy laparotomy scar gaping of 1.5×1.0 cm with granulation tissue in situ in periumbilical region. All haematological and biochemical investigations were within normal limits.

CLINICAL DIAGNOSIS

Sub-Acute Intestinal Obstruction

DIFFERENTIAL DIAGNOSIS

- Juvenile Xanthogranuloma (JXG),
- Extranodal Rosai-Dorfman Disease (RDD)
- Histiocytic Sarcoma, GI tract.

JXG is extremely rare and is usually found in the setting of systemic disease.[1]

PATHOLOGICAL DISCUSSION

Histopathological examination of small intestine revealed eosinophilic granuloma, Langerhans cell histiocytosis.

Langerhans cell histiocytosis (LCH) is a rare disease of unknown aetiology. LCH is characterized by the proliferation of bone-marrow-derived Langerhans cells.[2] Langerhans cells are a subtype of histiocyte, which in turn refers to large white blood cells (WBCs) present in tissues. In general, the term 'histiocyte' also includes macrophages and dendritic cells. The World Health Organization classification of haematopoietic and lymphoid tumours classified disorders of histiocytes into three specific categories, namely dendritic cell disorders, macrophage-related disorders, and malignant histiocytic disorders. LCH falls under the first category of dendritic cell disorders.[3]

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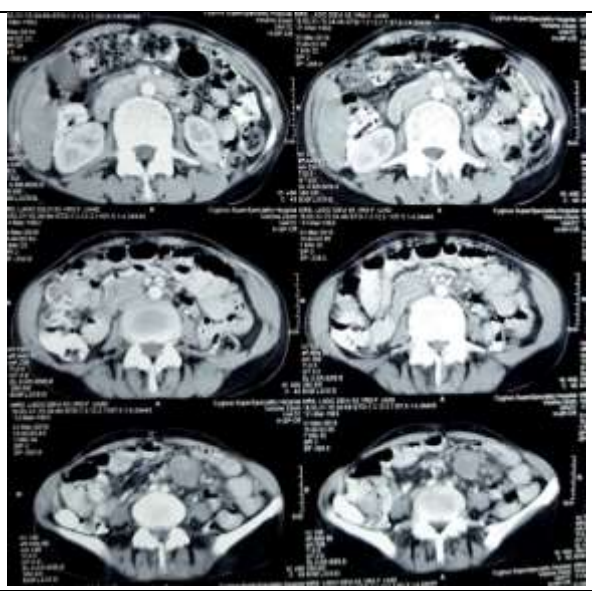

Figure 1. Axial CECT Abdomen- Serial Images showing Intestinal Obstruction

In mid twentieth century, Lichtenstein and Jaffe, described it as eosinophilic granuloma. They subsequently found that eosinophilic granuloma, Hand-Schuller-Christian disease and Letterer-Siwe disease represent manifestations of a single entity and termed it Histiocytosis X. In 1985, Langerhans cell histiocytosis (LCH) term was coined to encompass the entities which were formerly known as Histiocytosis X.[4]

It is more common in children and males as compared to adults and females with peak age of incidence being less than 2-years.[5] In LCH gastrointestinal (GI) tract involvement is exceedingly rare with only 10 cases of GI-LCH reported so far in adults.[1,6] Further in the GIT, LCH involvement of the stomach, small intestine, colon and perianal skin has been reported.[7] From the histopathological viewpoint, the demonstration of LC (Birbeck) granules by electron microscopy remains the "gold standard" for diagnosis of the phenotype, but expression of the CD1a antigen on lesional cells also provides the basis for definitive diagnosis.[8]

Typically, intestinal LCH is part of a wide-spread severe disease and has dismal prognosis. Although GI involvement is not a criterion for poor prognosis but literature review shows that more than half of the patients of GI-LCH die within first two years of diagnosis.[5]

DISCUSSION OF MANAGEMENT

With the diagnosis of Langerhans cell histiocytosis of small intestine and keeping in view of the poor general condition, she was started on chemotherapeutic trial with cyclophosphamide 50 mg orally twice daily. Patient received the above therapy for 2-months and was lost to follow-up thereafter.

LCH has been clinically classified into three major groups: unifocal multifocal involvement of a single organ system and multisystem LCH (where two or more organ systems are affected).[2] This classification is the basis of risk stratification to determine an appropriate chemotherapeutic regimen. Vinblastine, prednisolone, and/or etoposide are effective as first-line treatment for LCH.[9] Monotherapy with 2-Chlorodeoxyadenosine (2-CDA) is an effective second-line treatment in those who fail first-line therapy.[10] Promising results in the treatment of patients with severe, resistant to conventional therapy multi-system- LCH have been reported by a regimen using combination of 2-CDA and cytarabine.[11] Another option for such patients is haematopoietic stem cell therapy. In our patient, due to old age and low performance status, we put her on oral metronomic chemotherapeutic trial with tab cyclophosphamide chemotherapy but she subsequently lost to follow-up.

LCH involving the GI tract in adults is very rare and usually presents as an incidental finding. In adults with nonspecific GI symptoms it should be included as a differential diagnosis. Biopsy is essential to confirm the diagnosis of LCH with additional investigations to rule out systemic disease. Owing to the rarity and an incomplete understanding of the pathogenesis, treatment and chemotherapy regimens are not standardized. Due to the lack of data available to guide the treatment of LCH, the course of disease and prognosis vary on a case-by-case basis. Generally, patients with a multifocal and multisystem disease face unfavourable outcomes like in our patient.

FINAL DIAGNOSIS

Langerhans's Cell Histiocytosis of Small Intestine.

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