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Malignant Histiocytosis Associated with Malabsorption*

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Although malabsorption associated with malignant lymphoma has been recognised since 1937,[1] only recently has an association of malabsorption with malignant histiocytosis (MH) in adults has been published.[2]

In this report, we present a rare complication of MH in the pediatric age group.

Case Report

A 14 year old girl was admitted to Hacettepe Children's Medical Center for evaluation of intractable diarrhea. Three months prior to admission she had developed fever, diarrhea, vomiting spells and had felt pain around the umbilicus, all of which had been unresponsive to treatment. Her medical history indicated that she had been entirely well and her family history was unremarkable. With the exception of the chronically ill appearance, physical examination did not reveal any abnormalities.

Laboratory data on admission included a hemoglobin level of 12.3 gm/dl and white blood cell counts of 3800/mm^3 with 60 % segmented cells, 34 % lymphocytes and 6 % eosinophils. Results of urinalysis, chest X-ray and gastrointestinal series including pancreatic angiography were normal. Examination of stool specimens for ova and para-

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sites gave negative results as did bacterial and viral cultures. Immunoelectrophoresis disclosed no abnormality. Peroral jejunal biopsy revealed the expected amount of plasma cells and lymphocytes in the lamina propria but the surface epithelium was not preserved. One month following admission to the hospital, she developed anemia, leukopenia and thrombocytopenia. However, bone marrow aspiration revealed normal myeloid and erythroid cells. Three months after hospitalization meningeal signs appeared but cerebro-spinal fluid values (cell counts, protein, sugar) were within normal limits. During the 4 months hospitalization period diarrhea continued, with 10-15 episodes daily and recurrent pyrexial state, general weakness and anorexia. Liver and spleen enlargement were never noticed during this period and lymph nodes were not palpable. Pancytopenia remained severe throughout hospitalization. Repeat bone marrow aspiration disclosed hypercellularity with preponderance of monocytic and promyelocytic cells and abnormal histiocytes with abundant foamy and vacuolated cytoplasm containing phagocytosed erythrocytes, leukocytes and platelets. The diagnosis of MH was reached on the basis of final bone marrow findings and clinical features. She died 2 weeks later in spite of intensive chemotherapy.

Postmortem examination revealed diffuse infiltration of abnormal phagocytic histiocytes in lymph nodes, liver, spleen, kidney, pancreas thyroid, myocardium and meninges. Some of the organs were infected with candida albicans.

Discussion

The term MH is used by Rappaport[3] synonymously to histiocytic medullary reticulosis which was first used by Bodley-Scott and Robb-Smith in 1939.[4] The disease is generally associated with rapid weight loss, fever, lymphadenopathy, hepatosplenomegaly which is often accompanied by progressive anemia, leukopenia and thrombocytopenia. Malignancy associated with malabsorption is generally seen in Hodgkin's disease,[5] reticulum cell sarcoma[6] and mediterranean lymphoma which is the focus of increasing interest.[7] This condition has also been reported, however, rarely, in patients with juvenile chronic myelogenous leukemia.[8,9] Recently, its infrequent relationship with MH has been indicated by Isaacson and Wright.[2] In this case, malabsorption and fever of unknown origin were the only clinical manifestations. The most common clinical features of malignant histiocytosis were not obvious. Bone marrow aspiration finally revealing an increased number of phagocytic histiocytes, which appears to be a late event in the course of the disease,[10] established the diagnosis. This was confirmed by necropsy findings. No

intestinal infiltration was observed and therefore, diffuse pancreatic infiltration was assumed to be the major cause of malabsorption. Although, the clinico-pathological manifestations of the disease are well known, most of the MH cases have only been diagnosed at post-mortem. Awareness of possible association of malabsorption with MH and careful bone marrow examination in suspected patients, as in the case presented might provide for earlier diagnosis and more effective therapy.

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