

Letterer-Siwe Disease

Letterer-Siwe disease is the least common of the histiocytoses, comprising about ten per cent of all histiocytic disorders. It is a progressive, acute syndrome in children under three years of age, involving multiple visceral organ systems, such as the liver, spleen, lymph nodes and skin, and associated with purpura, bleeding gums, and multiple lesions similar to those seen in Hand-Schüller-Christian disease. The skull and pelvis are frequently involved. The skeletal lesions tend to be more diffuse than with the other histiocytoses and appear similar to that of leukemia or diffuse lymphoma. These patients usually die of bacterial infections within one or two years of their acute clinical onset because of bone marrow suppression. Histologically, the lesions look very similar to eosinophilic granuloma or Hand-Schüller-Christian disease, although it is unusual to see foam cells in this form of histiocytosis. Treatment consists of chemotherapeutic agents similar to those used in leukemia.