

Hand-Schüller-Christian Disease

Hand-Schüller-Christian disease is the intermediate form of histiocytoses-X that affects predominantly children, two-thirds of the cases being younger than five years of age. The classic triad for this syndrome is diabetes insipidus, exophthalmos, and single, geographic lesions involving mostly the skull and pelvic structures. The initial lesions appear like eosinophilic granuloma and, in fact, eosinophilic granuloma can progress into a Hand-Schüller-Christian type syndrome as the disease advances. It is common to have soft tissue involvement of lymph nodes, liver, spleen, lung, brain and kidney, as well as the skin changes that are also be seen in eosinophilic granuloma. Histologically, the same histiocytic cells as are seen with eosinophilic granuloma are present, along with eosinophils. In the later stages, foam cells and cholesterol deposits are typical. As the disease progresses and more and more soft tissue organs are affected, the prognosis worsens with an overall fatality rate of 10-30 per cent. Treatment consists of local resection plus systemic treatment with chemotherapeutic protocols similar to those used with leukemic patients.