

Eosinophilic Granuloma (Langerhans Histiocytosis)

The so-called benign histiocytoses, sometimes referred to as histiocytosis X, include various disease conditions such as eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease. Eosinophilic granuloma is the most benign of the histiocytic disorders. Hand-Schüller-Christian disease presents with an intermediate diffuse process of both bone and soft tissue and can be fatal. Letterer-Siwe disease is the most aggressive and fatal form of the histiocytoses, presenting like leukemia with a very poor prognosis for survival.

Eosinophilic granuloma, now referred to as Langerhans histiocytosis, is a benign histiocytic disorder presenting in children between the ages of 5 and 15 years with a clinical picture that can masquerade as a malignant neoplasm such as Ewing's sarcoma. It occurs twice as often in males as females. It is usually a monostotic disorder of the skeletal system, however in ten per cent of cases it will be seen in two or three separate skeletal sites. The etiology of this histiocytic process is still unknown but some have postulated a viral etiology. Patients present with inflammatory pain that is more severe at night and may be associated with a low-grade fever or elevated sedimentation rate. The most common location is in the skull, followed next by the ribs, pelvis, maxilla, vertebral body, clavicle, and scapular, in that order. Besides flat bone involvement, it is commonly seen in diaphyses of long bones where it can masquerade as Ewing's sarcoma, but it can also occur in metaphyseal bone. The least common location is in epiphyseal bone. In young children, the condition can be extremely permeative and destructive in nature, taking on the appearance of Ewing's sarcoma, metastatic neuroblastoma, or acute osteomyelitis.

On X-ray, eosinophilic granuloma has an onionskin appearance similar to a Ewing's sarcoma. In an older age group, the condition tends to be more focal and more granulomatous in appearance with less permeative change. Microscopically, there are large, pale-staining histiocytes speckled with small, bright-staining eosinophils and an occasional giant cell. Eosinophilic granuloma tends to involute spontaneously without treatment and therefore symptomatic treatment should be conservative -- simple curettage for diagnostic purposes and perhaps cortical steroid injections to inhibit the inflammatory process. In the parts of the body more difficult to access, such as the spine or pelvis, very low-grade radiation therapy can be considered, realizing that this could convert the process to a malignant sarcoma as a later date. A low dosage chemotherapy program can be considered with the more aggressive forms of eosinophilic granuloma, with multi-focal involvement, and especially if there is soft tissue

involvement of the skin, lymph nodes or lung. Sometimes the low-grade eosinophilic granuloma histiocytosis can upgrade to a more aggressive and dangerous form such as Hand-Schüller-Christian disease or even Letterer-Siwe disease. With spinal lesions, spinal cord compression can result in paraparesis requiring laminectomy decompression. However, kyphotic deformities in younger patients tend to correct spontaneously without anterior spinal instrumentation.