Histiocytosis is a group of diseases for which proliferative process in monocytic-macrophagal system is typical. There are various forms of the diseases with their peculiarities of clinical manifestations, prognosis and treatment depending upon degree of maturity and differentiating of histiocytic elements.

Epidermal cells with multiple pseudopodial branching on their surface are called Lanherhans cells; they were found in disease known as “Histiocytosis X” the name of which was changed for “Histiocytosis of Langerhans cells” in 1987. Immunopathological and tumorous origin of the disease is under discussion. High frequency of spontaneous remissions, low mortality rate and absence of chromosomal anomalies in the cells of foci of affection testify to immunopathologic origin of the disease, clonal character of Langerhans cells proliferation – to tumorous origin. Cytokines are instrumental in pathogenesis of the disease. Morbidity is 2 to 10 cases upon 1 000 000 of children below 1 years. Middle age is 30 months but age of the patients may vary from birth up to 90 years.

From 2005 up to 2015 there were 10 children under our supervision aged from 9 months up to 17 years, among them 9 boys and 1 girl. All of them suffered from eosinophilic granuloma.

Clinical manifestations varied from single focus of affection up to progressive disseminated forms which led to multiorganic insufficiency. Marked early clinical manifestations included fever (80%), hepatomegaly (60%), splenomegaly (10%), neurological symptoms (20%), rash (10%) and lymphadenopathy (10%).

Single or multiple affections of bone were detected most frequently: skull (60%), spine (50%), bones of extremities (20%). Affection of the bone manifested with pain and tumor from the focus of destruction. Retro-orbital localization of pathologic foci provoked exophtalm (20%). One child had seborrheic rash on the head skin which was diagnosed as seborrheic dermatitis. One child more have had red papulous rash in the groin and on the abdomen which looked like diffuse fungal affection of skin.
Hypothalamic dysfunction manifested with obesity and affection of the posterior hypophysial part (neurohypophysis) provoking diabetes insipidus was seen in 10% of cases.

Laboratory examinations included CBC + hematocrit + platelets, functional liver test, proteinogram, coagulogram, X-ray of the chest, ultrasound examination of inner organs, CT scan, PET, MRI, puncture and biopsy of bone marrow, determination of pulmonary function, examination of small intestine with biopsy (if indicate), consultations of surgeon, ophthalmologist, neurologist and ENT-doctor.

Differential diagnosis has been made with seborrheic dermatitis, osteomyelitis, tuberculosis of bones, osteosarcoma, neuroblastoma, fibrous osteodystrophy, lymphoma, leucosis, and lymphogranulomatosis.

Own observation of eosinophilic granuloma is described. The disease started with stomatological problems; diagnosis of adamantinoma of mandible was made. The child was examined by endocrinologist and gastroenterologist; diabetes insipidus was diagnosed. Later the child was examined by neurosurgeon; diagnosis of hamartoma of hypothalamic-hypophysial zone was made. Correct diagnosis of eosinophilic granuloma has been made almost in four years after the beginning of the disease after performing of histological examination. Diagnostic process in a child was long and difficult due to various and entangled clinical manifestations which camouflaged the main problem and delayed correct diagnosis.

It is concluded that histiocytosis is not frequent but severe disease that demands careful differential diagnostics. If diagnostics and treatment are made in time survival is rather high.