Irradiation of Histiocytosis X Confined to the Oral Mucosa

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Background: Histiocytosis X is a rare granulomatous disease and it is still under debate as to whether this disease has to be counted among malignant tumors or benign immunologic lesions. The typical localization is confined to the bone. In rare cases histiocytosis X may be localized in the oropharyngeal mucosa.

Patients and Methods: We report about a case of a 75-year-old woman presenting with histiocytosis X lesions solely located in the oral cavity. A total dose of 19.5 Gy was administered without any side effects.

Results: Two weeks after completion of irradiation a rapid regression of mucosal ulcerations and improvement of symptoms occurred. In a follow-up of 24 months the patient is still in complete remission observed.

Conclusion: Review of the literature revealed only casuistic reports about solitary lesions of histiocytosis X without bone involvement in the adulthood. External beam radiotherapy with total doses between 10 and 20 Gy seems to be effective for local control of this disease.

Key Words: Histiocytosis X · Solitary lesion · Radiotherapy

Introduction
Histiocytosis X encompasses a distinct group of disorders of the reticuloendothelial system that have different clinical features but share common histological characteristics, including abnormal proliferation of Langerhans cells. The etiology remains unexplained and is attributed so far to undefined immunologic disturbances. Prognosis of the different types depends on the extension of the disease. It is still under discussion as to whether the disease represents a malignant tumor or is just a manifestation of benign immunologic disorders [7].

Localized forms of histiocytosis typically affect a single or several bones. In selected cases lesions can also be localized in the oropharyngeal mucosa [9].

We report about such a case with histiocytosis X exclusively located in the oral cavity in an adult patient. The patient was successfully treated with radiation.

Case Report
A 75-year-old woman initially presented with localized mucosal lesions in the right oral cavity. The patient complained...
about pain and moderate dysphagia but did not report any further symptoms. Because of ineffective local antibiotic and steroid treatment and rapid involvement of the whole buccal gingiva, 2 biopsies were performed revealing diagnosis of Langerhans cell histiocytosis with typical histologic features.

The patient underwent clinical and radiological examinations which showed multiple ulcerated lesions and edema of the whole oral mucosa (Figure 1). Computed tomography scan of the head revealed no lytic lesions. Chest X-ray, thoracic and abdominal computed tomography, bone scan and pathologic examination of the bone marrow showed also no further manifestations of the disease.

A total dose of 12 Gy was administered in conventional fractionation over 10 days through 2 lateral wedged fields. After 2 weeks the mucosal lesions appeared to have blanched, but had not regressed. Therefore irradiation was continued to a total dose of 19.5 Gy, which was tolerated without any side effects. Two weeks after completion of radiotherapy regression was almost complete (Figure 2) and the patient became symptom-free. A rapid improvement of mucosal ulcerations could be observed and 2 months later clinical examination revealed an almost complete remission. The patient is now 24 months after radiotherapy and upon latest follow-up no disease progression was observed.

Discussion

Histiocytosis X represents a rare granulomatous disease, of which etiology and pathogenesis is still unknown and that usually occurs in children with a mean age of 15 years [2].

The histological features are characterized by idopathic proliferation of Langerhans cells or their marrow precursors; somatic mutation of DNA of Langerhans precursors are reported and hypothesis of typical deletions of tumor suppressor genes is still discussed. Up to now there is no clear distinction if histiocytosis X is a primary malignancy or reactive immunologic disorder. In rare cases related disorders like benign variants as self-limited eosinophilic ulcerations could be detected [1, 11]. In most cases the disease is confined to the bone, most frequently located in the skull and femur [3].

Prognostic factors are number of involved organs, age and the presence of anemia, thrombocytopenia or spleen involvement. The favorable risk group is characterized by unifocal lesions, in which minimal therapy is often sufficient for sustained control. Dissemination is found to be more likely in males than in females with other unfavorable parameters such as lesions of the liver and lung or age under 2 years. In disseminated disease also cerebral involvement has been described. Fatal cases are always associated with multisystem disease [5, 6]. In a study of 155 children 6-year overall survival ranged between 48 and 90% depending on the patient’s age and organ dysfunction [7].

Often, the clinical behavior is similar to common diseases such as dermatitis, otitis, mastoiditis or gingivitis as shown in our case.

The therapeutic procedures of histiocytosis X include surgery, radiotherapy and chemotherapy. Extensive disease of Langerhans cell histiocytosis is usually treated by chemotherapy with vincristine and prednisolone [9]. For localized disease general treatment is surgery, postoperative radiotherapy is applied in selected cases [2]. Solitary histiocytosis X lesions appear to respond favorably to radiotherapy, however, irradiation in childhood is hampered by a small but definite risk of side effects, especially disruption of growth pattern with dysmorphic or retarded growth and secondary malignancies [5, 12]. Therefore the major indication for irradiation in