

Severe oral involvement in a case of Hand-Schuller-Christian disease

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SUMMARY

We report the case of a patient with diabetes insipidus who came to our observation because of severe lesions of the oral mucosa. He referred that he had lost all his molars and three incisors over the past two years. The clinical cutaneous examination revealed the presence of small yellowish-brown papules on the forehead only. A biopsy performed from the gingival mucosa showed the presence of Langerhans cells and a dense inflammatory infiltrate. The immunohistochemical examination showed CD1 antigenic determinant and S-100 protein positive. Considering the presence of diabetes insipidus, the clinical, histological, immunohistochemical picture, the diagnosis was of a Hand-Schuller-Christian disease. The patient underwent chemotherapy and radiotherapy. Three months after the end of the therapy a gingival biopsy confirmed the success of the treatment.

KEY WORDS

Hand-Schuller-Christian disease, Langerhans cell, oral involvement, histiocytic disorders

Introduction

Histiocytic disorders are a group of heterogeneous diseases resulting from the proliferation and dissemination of pathologic histiocytic cells or Langerhans-like cells producing focal, localised single form or disseminated multisystem manifestations. A correct classification mainly depends on the histological aspects.

Case report

B.G. came to our observation because of the worsening of clinical manifestations of oral mucosa. For

about ten years the patient was suffering from idiopathic diabetes insipidus and had been treated with desmopressin nasal spray.

He reported a history of recurrent gingivitis since 1991 with halitosis, teeth unsteadiness, jaw swelling, periodontal lesions, haemorrhages and necrotic lesions that seriously destroyed his gums. He had lost all his molars and three incisors over the past two years. During the oral examination, made difficult by the limited opening of the mouth, it was possible to observe that the tissue of both the upper and lower gums appeared swollen, intensely red in colour and prone to bleeding.

