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 Langherans 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-Shuller-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 Langherans-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.

It was also possible to observe serious unsteadiness of all the other teeth (Fig. 1). We took a panoramic radiograph of both dental arches, which showed a remarkable atrophy of the alveolar ridge and a severe parodontitis. The remaining teeth were abnormally sited in an extra alveolar position (Fig. 2). The vertebral X-rays showed arthopathic changes that have spread mainly at cervical level. Thoracic radiography, liver and spleen ecography didn't show any changes. The clinical cutaneous examination revealed the presence of small yellowish-brown papules on the forehead. The patient reported that the periodic appearance of these lesions were out of the blue and, in his opinion, unrelated to any other symptoms. The lymphonodal apparatus was normal.

A biopsy performed from the gingival mucous showed the presence of Langherans cells in a dense inflammatory infiltrate. The immunohistochemical examination showed CD1 antigenic determinant and S-100 protein positive (Fig. 3).

Considering the presence of diabetes insipidus, the histological and immunohistochemical data, the chronic evolution of the clinical picture the diagnosis was of a Hand-Shuller-Christian disease.

Later the scintigraphic examination of the skull was performed on the patient. There was no evidence of any other localised bone lesions.

The patient underwent chemotherapy (a cycle of vinblastine -0.2 mg/kg once a week- plus prednisone - 30 mg/day) and radiotherapy (radiation doses ranged from 800-1500 rad - 8-15 Gy - with a mean dose of 1066 rad - 10.66Gy).

Three months after the end of the therapy a gingival biopsy confirmed the success of the combined treatment. Five years after the diagnosis was made, the patient is symptom-free.

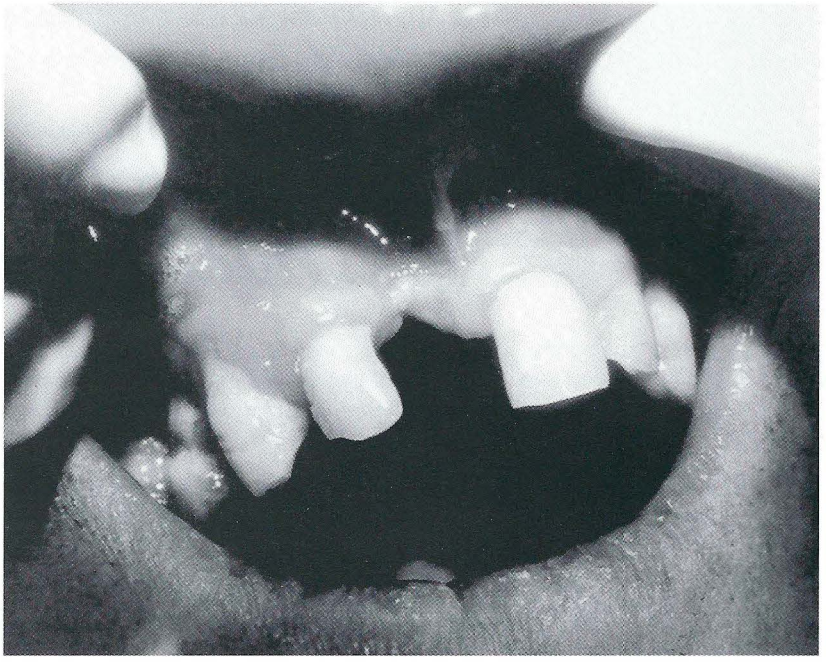


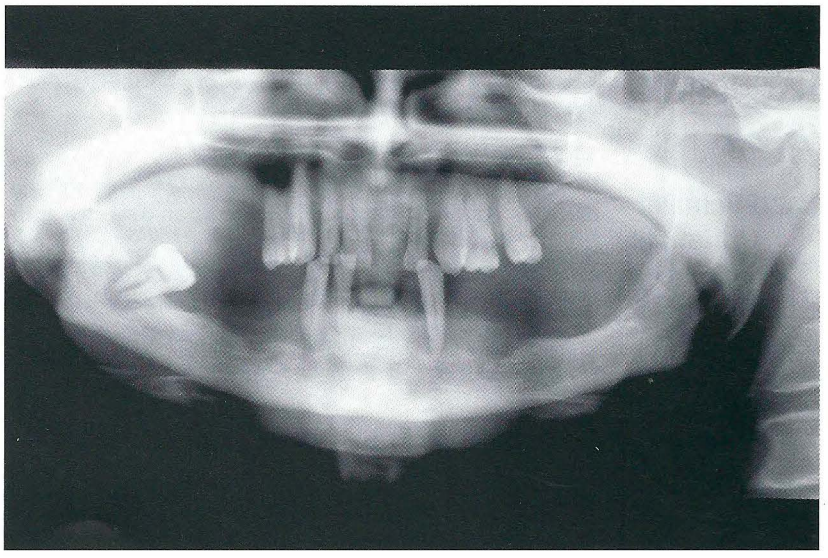
Figure 1. Oral manifestations in the patient.

Discussion

Histiocytic disorders are a group of heterogeneous diseases resulting from the proliferation and dissemination of pathologic histiocytic cells or Langherans like cells producing focal, localised single form or disseminated, multisystem manifestations (1). A correct classification mainly depends on the histological examination, ultrastructural and immunocytochemical data (2).

As far as Langherans cell histiocytosis (LCH) is concerned (Hand Shuller Christian disease, Abt Letterer Siwe disease, Eosinophilic granuloma), the diagnosis is suspected on the clinical presentation. Distinct lesions are usually seen in soft tissue and bone; lungs and skin often show a more diffuse, patchy involvement; the lesions are soft and yellow to brown with frequent areas of necrosis and haemorrhage. Histologically, LCH exhibits hyperplasia and proliferation of the reticuloendothelial system, infiltration with CD1 positive histio-

Figure 2. Panoramic radiographs showing extensive alveolar bone destruction.



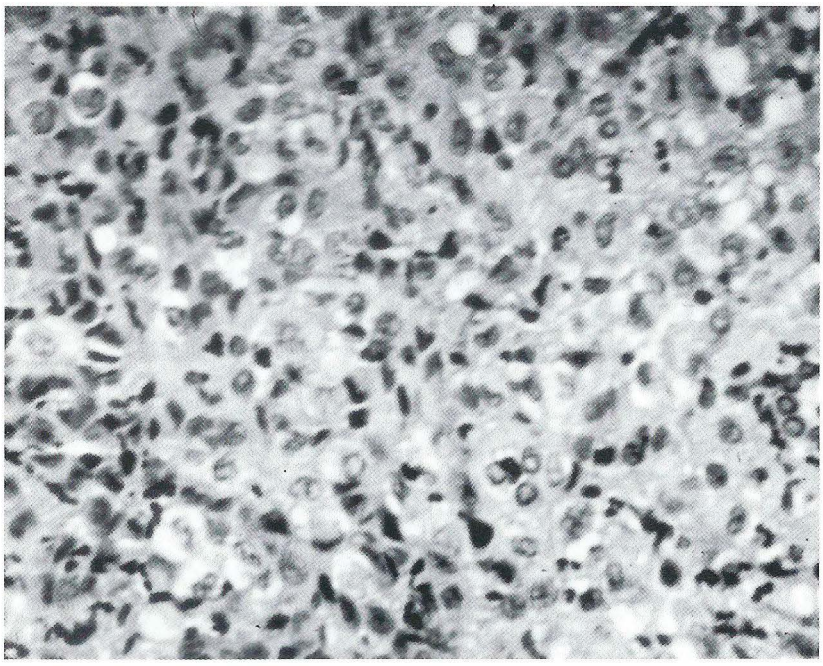


Figure 3. Langerhans cells agglomerations mixed with inflammatory elements (hematoxylin - eosin, magnification x 40)

cytes disclosing intracytoplasmatic Birbek granules at the electron microscopic examination (3).

Causes and pathogenesis of the disease remain unclear. However, recent studies suggest a disorder of the immune system as an important factor in the aetiology of LCH (4,5,6).

The prognosis depends on the patient's age at the onset and the extent of the disease (7,8). Treatment can be separated into management of solitary symptomatic lesions and that of systemic disease (9).

Among LCH, Hand-Shuller-Christian disease (15-40% of cases) is the chronic form clinically characterised by a triad in which characteristic multifocal bone lesions and extra-skeletal involvement of the reticulo-endotelial system are combined with exophtalmus in about 10% of cases and diabetes insipidus (6,10,11,12). The course is extremely variable: often chronic, seldom fatal (13). The histological diagnosis is based on the presence of a histiocytic infiltrate in the upper and middle dermis. In particular, the optical microscope examination reveals in the papillary dermis an important oedema, large cells with a reniform or indented nucleus and abundant eosinophilic cytoplasm. Histochemical colouring, showing positiveness for S-100 protein, the presence of CD1, CD4 and HLA-DR surface antigens confirm the diagnosis (12,14,15).

Conclusion

The therapy of the Hand-Schuller-Christian disease varies according to the age of the patient, the severity and extent of the clinical picture. It includes an orthopaedic-surgical approach, radiotherapy, chemotherapy and/or immunotherapeutic treatment (13).

We report this case because of diagnostically important lesions of the oral mucosa in a patient with diabetes insipidus without exoftalmus and without evident cutaneous lesions.

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