

Extensive Basal Cell Carcinoma and Disseminated Lesions in Hand-Schüller-Christian Disease

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Sir,

Langerhans' cell histiocytosis (LCH) includes four clinical entities with a common characteristic of infiltration of LC in different tissues. Biopsy of suspicious lesions and staining for CD1a and S100 protein or antilangerin are needed to establish a definitive diagnosis of LCH. Electron microscopy to identify Birbeck granules is performed less frequently because of time and cost constraints (1, 2).

The pathogenesis remains unclear. A neoplastic aetiology has been suggested due to the existence of a single clone in the infiltration, although no cytogenetic abnormalities have been found (3, 4). Most investigators consider LCH to result from an immunological dysfunction (2, 5, 6). The presence of aggregates of other immunologically active cells, decreased numbers of suppressor T lymphocytes and high levels of cytokines in the lesions suggest an exaggerated physiological response of LC to an antigen, or an appropriate response to abnormal signals from other immune cells (1, 7, 8). Although numerous facts are known about the cell mediators, it has not been possible to prove the

participation of any factor in the unchaining of the clonal proliferation of the cells in the LCH (9).

We present here a new case of basal cell carcinoma (BCC) in association with one type of LCH, Hand-Schüller-Christian disease, with a probably late specific cutaneous infiltration of the LC.

CASE REPORT

A 41-year-old man presented with a 3-year history of an extensive supra-auricular tumour and was admitted for elective surgery in our department.

His past medical history included Hand-Schüller-Christian disease with multi-bone involvement diagnosed at the age of 5 years and treated with radiation, systemic chemotherapy (prednisone, vincristine, chlorambucil, cyclophosphamide) and curettage.

A general physical examination revealed deafness, some orthopaedic defects, and a characteristic face with a wide forehead, a sharp nose and a prominent lower jaw with missing teeth. The cutaneous examination showed a 5 × 2 cm ulcerated and crusted plaque with ill-defined margins in the right supra-auricular region (Fig. 1). No palpable regional lymph nodes were present. Hyperkeratotic, pearly and clustered papules on the scalp, frontal, mandibular, retroauricular and occipital regions were also noted. The presternal region and the anterior aspect of the arms demonstrated similar lesions. These lesions were asymptomatic and had gradually developed over recent years, but the patient was not concerned enough to seek medical evaluation. He also presented verrucous plaques on the lateral aspect of the fingers, suggestive of warts, that had been resistant to multiple therapies.

A wide surgical excision of the right auricular area was performed. The involved auricular cartilaginous tissue was removed by Mohs surgery and the defect was closed with a free total skin graft of the abdominal region, with an excellent aesthetic and functional result (Fig. 2). The histological study showed a solid BCC.

The patient refused to undergo a cutaneous biopsy of the lesions of the face and the trunk to confirm the diagnosis. At 8-month follow-up examination, the patient was in good health, without evidence of recurrence of the skin tumour. A few months later, the patient died of bronchopneumonia. No autopsy was performed.



Fig. 1. Extensive basal cell carcinoma on the right auricular region.

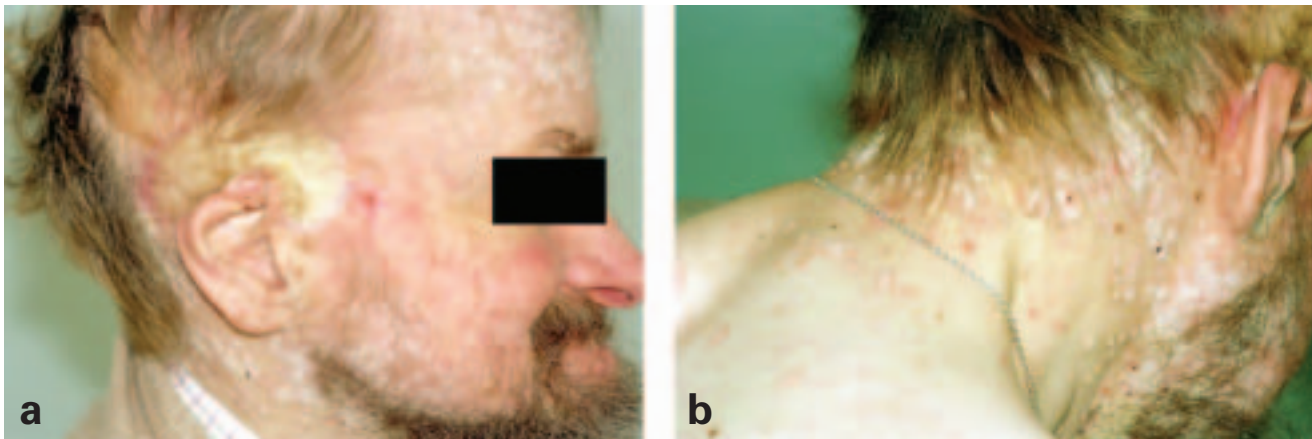


Fig. 2. (a) Hyperkeratotic papules on the frontal, mandibular and retro-auricular regions and graft at 4 months after surgery. (b) Detail of the pearled papules on the scalp and the occipital region.

DISCUSSION

Associations between malignant neoplasms and LCH have been described in several series, possibly favoured because of the alteration of the humoral immunity. These malignant neoplasms include lymphoma, leukaemia, lung carcinoma, thyroid carcinoma, breast carcinoma, astrocytoma, glioblastoma, medulloblastoma, ependymoma, retinoblastoma, hepatocellular carcinoma, gastric carcinoma, Ewing sarcoma, tongue carcinoma and osteosarcoma. In general, the intimate and simultaneous association of LCH with malignant lymphoma and lung carcinomas suggests strongly that the process that leads to the association is a reactive one. However, in the patients with acute non-lymphoblastic leukaemia and other solid tumours, the latency of the malignant neoplasm after the diagnosis of LCH suggests a causal relationship to therapy (chemotherapy and radiation therapy) used in the treatment of LCH (2, 9–11).

Our patient presented with an extensive BCC in the auricular region. The family history was negative for cutaneous carcinoma, and the sun exposure had been very limited. Only four similar cases have been reported previously in the literature (10, 11). All patients developed a BCC in a previously irradiated area, with a median of 12.5 years after LCH diagnosis. Probably the multiple treatments with radiation therapy and systemic chemotherapy in infancy were responsible for the increased risk of secondary malignant neoplasms in all cases.

Our hypothesis is that the cutaneous lesions located mainly in seborrhoeic areas could represent a specific cutaneous infiltration of LC. This could not be demonstrated as we were unable to obtain skin biopsy. The late cutaneous infiltration is a fact scarcely reflected in the literature. The patient's lesions were highly

suggestive of a specific infiltration, because of their morphology and distribution.

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