Merkel-cell Carcinoma in Behçet's Disease

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Sir,

Merkel-cell carcinoma (MCC) is a rare malignant tumour of the skin which is highly aggressive; frequent recurrences and distant spread are common (1). Behçet's disease (BD) is a multisystem vasculitis, with oral and genital aphthous ulcers, cutaneous vasculitis, uveitis and arthritis as its main features (2). Association of BD with malignant tumours, mainly solid neoplasms, has been described, but a review of the literature showed no report of an association between BD and cutaneous tumours. We herein report the case of a patient affected with BD who subsequently developed a MCC.

CASE REPORT

A 67-year-old man had suffered from systemic BD for 8 years, particularly involving the eyes. He had had one episode of deep venous thrombosis and many episodes of pneumonia. The patient was treated for 6 years with immunosuppressive therapy, viz. cyclosporine 150 mg 6 days a week, methotrexate 10 mg plus methylprednisolone 80 mg a week, cyclophosphamide 400 mg plus methylprednisolone 80 mg a week. About 6 years after the start of therapy it was noted that the skin of the middle third of his left forearm showed a well defined, oedematous-erythematous infiltrating plaque, with an irregular surface, 85 mm $\times$ 120 mm in size, that had appeared 3 months before. The histological examination was consistent with the diagnosis of MCC. Immunohistochemical analysis showed that the neoplastic cells were positive for synaptophisin, neuron-specific enolase and perinuclear cytokeratin 20, which are all markers consistent with the diagnosis of MCC (1). Instrumental staging for MCC, including chest X-rays, abdominal and lymph node echography, and total body tomographic scan, did not show any distant spread. The patient was sent to an oncologic unit, and the immunosuppressive therapy was stopped except for cyclosporine; instead, radiotherapy was carried out, followed by chemotherapy. Unfortunately, the patient died 7 months after discontinuation of immunosuppressive treatment.

DISCUSSION

To the best of our knowledge, only 43 cases of BD associated with malignant diseases have been described up to now, some lymphoid/haematological (18 cases) and others solid (25 cases) but no cutaneous neoplasms. In particular, BD has never before been reported in association with a MCC. However as BD is now recognized not only as recurrent mucous aphthae and relapsing uveitis (3), but also as a multisystemic vasculitis (2), a risk of association with cancer is not unexpected (4–6), although rare (7).

In several instances, the association of BD with solid tumours was considered as incidental (7). However, we believe that immunosuppression for BD might trigger MCC. In fact, 16 MCC cases have been reported arising after iatrogenic immunosuppression (mainly given for renal transplantion) (8–10). Thus MCC should be kept in mind when evaluating an atypical skin lesion in the context of iatrogenic immunosuppression, for example in patients with BD.

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