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PhotoDermDiagnosis

Nodular eruption on the back of scalp

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A 60-year-old male presented to OPD with itchy skin-coloured papules on the dorsum of hands, feet, elbows and lower half of legs followed by painful nodular eruption on the back of the scalp for the last 3 months. Patient gave history of weight loss and lumps on either side of neck and right groin. Rest of the systemic enquiry was unremarkable except that he was a chronic smoker.

Examination revealed erythematous polypoid outgrowths on the back of the scalp, a few of them were fused to form 8x6 cm mass, soft to firm (**Figure 1**). Two nodules were discrete 2x2 cm in size. Skin-coloured papules were present on the dorsum of hands, feet, elbows and legs with excoriation marks. Posterior cervical lymph nodes were enlarged bilaterally, they were firm, mobile and non-tender. Rest of the examination was unremarkable. Histopathology of the nodular lesion revealed stratified squamous epithelium, within the dermis there was collection of cells with hyperchromatic nuclei with prominent nucleoli (**Figure 2**). Immunostaining revealed LCA, CD30 and CD3 Positive while ALK was negative.

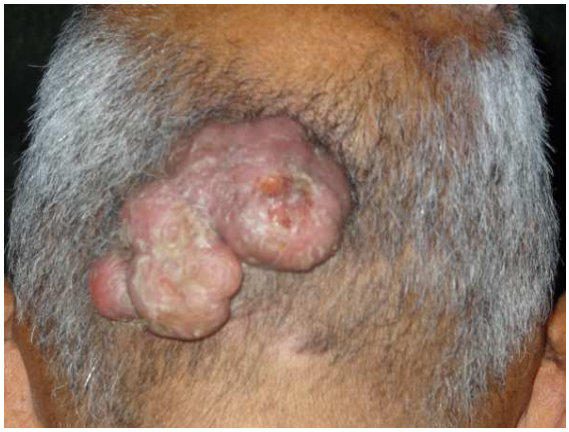

Figure 1 Polypoid outgrowths on the back of the scalp.

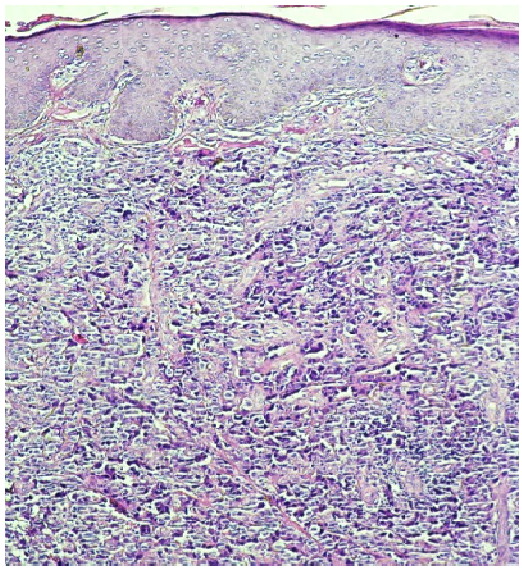

Figure 2 Stratified squamous epithelium, collection lymphocytes with hyperchromatic nuclei and prominent nucleoli in the dermis.

What is your diagnosis?

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Diagnosis

Non-Hodgkin's anaplastic lymphoma

Discussion

Anaplastic large cell lymphoma (ALCL) is a rare type of Non-Hodgkin lymphoma. ALCL comprises about 2% of all NHLs and approximately 20% of all T cell lymphomas.[1] ALCL can initially appear either in the skin, lymph nodes, or in organs throughout the body. ALCL that appears in the skin is called primary cutaneous ALCL, which has a less aggressive disease course than the systemic types.[2]

The characteristic features of primary cutaneous ALCL include solitary or multiple erythematous nodules or tumours that appear on any part of the body, often grow very slowly, and may be present for a long time before being diagnosed. They have a tendency to ulcerate, and may itch. Approximately 10% primary cutaneous ALCL extends beyond the skin to lymph nodes or organs. If this occurs, it is usually treated like the systemic forms of ALCL.[3] The presence of the protein, 'CD30 antigen' on the surface of lymphoma cells is the hallmark of the disease.[4] Systemic ALCL which can affect all organs in the body is aggressive; and primary cutaneous ALCL which is confined to the skin is slow-growing.[5] There are two subtypes: ALK-negative and ALK-positive, depending on whether the lymphoma cells produce a protein called 'anaplastic large cell kinase' (ALK). Patients with ALK-positive ALCL are generally younger and respond better to treatment than those with ALK-negative ALCL.[6]

Primary cutaneous ALCL usually presents as a solitary skin nodule. In 20% of cases there may be multiple nodules. Involvement of lymph nodes draining the affected region does not usually lead to more widespread disease.[7] The nodules may regress spontaneously, but also tend to recur. Primary cutaneous ALCL presents in older age groups (median age 55 years), and is rare in children. It is 2-3 times more common in men than in women.[8] Systemic ALK-positive ALCL is more likely to affect children and young adults (median age 34), although there is a group who presents later in life. Patients with systemic ALK-negative ALCL present at a later age (median age 58 years). Systemic ALCL is slightly more common in men than in women. The causes are unknown. It is not due to infection and cannot be passed on to others.[8]

Primary cutaneous ALCL may go into spontaneous remission. However this is inevitably followed by a relapse.

Treatment of primary cutaneous ALCL depends on the extent of skin lesions. If the disease is confined to a single lesion or area, radiation therapy or surgical excision will result in complete remission in approximately 95% of patients. If there are multiple lesions or relapsed disease in the skin, radiation and systemic chemotherapy can eradicate the skin lesions but will not reduce the likelihood of new lesions developing. Although primary cutaneous ALCL tends to relapse in about 40% of cases, the long-term prognosis remains excellent as long as relapses are confined to the skin. At relapse, radiation, topical treatments, mild chemotherapies, biologic therapies, or excisions for small lesions have all been shown to be successful in controlling the disease.[9]

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