SKIN CANCER (OTHER THAN MELANOMA)

CASE OF ANAPLASTIC LARGE CELL LYMPHOMA IN ASSOCIATION WITH X-LINKED IMMUNODEFICIENCY

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Background: Anaplastic large cell lymphoma (ALCL) is a rare subtype of lymphoma first described in 1985 by Stein et al., consistently expressing surface antigen CD30. ALCL can be classified as either primary cutaneous or systemic.

Observation: A 71-year-old male presented with a rapidly enlarging and painful lesion on his right elbow over three months. The lesion appeared as a red nodule initially and treated as an abscess but failed to respond to oral antibiotics and began to ulcerate.

He had a chronic lymphopenia and history of recurrent infections with human papillomavirus and cytomegalovirus. He had been diagnosed to have combined X-linked immunodeficiency on IL2RG gene.

Clinical examination revealed a large (5x4cm) erythematous indurated plaque with areas of ulcerations over the right elbow. He was afebrile and no lymphadenopathy was found.

Incisional biopsy of the lesion showed a nodular infiltrate of the dermis composed of large pleomorphic cells which are highly proliferative with high Ki67 and strongly positive to CD30 and CD4. These cells also expressed an abnormal T-cell phenotype with loss of CD3, CD5 and CD7.

This finding was in keeping with CD30-positive T cell lympho-proliferative disorder and a diagnosis of primary cutaneous ALCL was made.

He completed a course of radiotherapy and some initial improvement was observed.

Key message: We presented a case with an unusual presentation of primary cutaneous ALCL in association with X-linked immunodeficiency. Treatment of primary cutaneous ALCL is either local excision or radiotherapy. Radiotherapy can also be an adjuvant treatment to local excision to prevent recurrence. In general, the cause of ALCL is unknown. However, there have been several reports of primary cutaneous ALCL in patients with Epstein-Barr virus and human immunodeficiency virus. The presence of on IL2RG gene is a new finding.

The clinical importance of this finding is worth exploring in the future.