SKIN CANCER (OTHER THAN MELANOMA)

A RARE CASE OF INTRAVASCULAR CYTOTOXIC T-CELL LYMPHOMA WITH AN UNUSUAL PRESENTATION

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Background: Intravascular lymphoma (IVL) is a rare and aggressive subtype of non-Hodgkin's lymphoma characterized by proliferation of neoplastic cells exclusively in the vascular lumina. IVLs are predominantly of B-cell phenotype, but occasional cases of T-cell lineage have been described. We report a case of intravascular cytotoxic T-cell lymphoma (IVCTL) occurring in an immunocompetent male with cutaneous, ocular and testis involvement.

Observation: A 63-year-old man, with no past medical history, presented with an 8 month-history of asymptomatic indurated erythematous patches of the legs and weight loss. Physical examination revealed extended livedo racemosa disposed symmetrically on the trunk, arms, and thighs, bilateral eye redness and swelling of the testes. The patient had a left hydrocele. The right epididymis was swollen with a sensitive induration of the head and tail. Ophthalmologic examination showed bilateral anterior uveitis consistent with tumoral pseudo-uveitis. There was no fever or lymphadenopathy. Laboratory examination revealed elevated LDH level (1882 U/L). Both thoracic-abdominal-pelvic CT and brain and spine MRI scans were normal. A biopsy of both the skin and the right epididymis was performed showing similar histological features: the vessels lumina contained many large cells with reduced cytoplasm and mitotic figures. In immunohistochemistry, tumor cells were CD3(+), CD56(+), granzyme B(+), CD5(-) and CD30(-). The diagnosis of IVCTL was made. Bone marrow biopsy showed no tumoral infiltration. A multidrug chemotherapy was initiated.

Key message: IVLs of T-cell lineage are very rarely reported with less than 28 cases in the literature. We report a unique case of IVL with a cytotoxic phenotype and an unusual clinical presentation: livedo racemosa and tumoral pseudo-uveitis. The treatment of testicular involvement could be challenging and may require surgery.