

A Case of Wiskott-Aldrich Syndrome with HPV16-associated Cutaneous Carcinoma

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RATIONALE:

Squamous cell and verrucous carcinomas have not been reported with Wiskott Aldrich Syndrome (WAS).

CASE:

AM is a 21 year-old male who was diagnosed with WAS at two weeks of age after presenting with petechiae and otitis media. A matched bone marrow transplant donor was unavailable; the patient's only sibling was also affected with WAS. The patient underwent splenectomy at age seven for bleeding complications. Supportive treatment included prophylactic antibiotics, topical anti-inflammatory medications, and monthly IVIG. Beginning at the age of twelve, the patient had recurrent episodes of tinea pedis, unguis incarnatus, verrucae, eczema and bacterial superinfection. Topical therapies included steroids, antibiotics as well as calcineurin inhibitors. At age 20, the patient developed a small ulceration on his right foot that developed into a vascular-type polyp, as well as a large polyp between the 3rd and 4th toes on his left foot that caused pain with ambulation.

RESULTS:

Pathology of shave biopsies revealed squamous cell carcinoma in situ in the right foot and verrucous carcinoma in the left. *In situ* hybridization studies identified HPV16 in the tissue biopsies.

CONCLUSIONS:

Lymphoreticular malignancies occur in 12 - 18% of patients and are the most common cause of death in patients with WAS who survive early childhood. Two cases of cutaneous lymphoma have been reported in association with WAS, but there are no previously reported cases of cutaneous carcinoma. The development of malignancies in patients with WAS is poorly understood, but may be related to defective immune surveillance associated with immunodeficiency. In the case of AM, the use of topical immunomodulators in the context of oncogenic HPV infection may also have played a role.