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Buschke-Ollendorff Syndrome

Buschke-Ollendorf syndrome (BOS) is an autosomal dominant genodermatosis associated with heterozygous loss-of-function germline mutations in the *LEMD3* gene. It is characterized by small, acquired, asymptomatic foci of osteosclerosis (osteopoikilosis [OPK]) along with connective tissue nevi. In this case series, Yadegari et al describe a father and son with nevus elasticus along with radiographic evidence of OPK. These clinical findings were diagnostic of BOS, but *LEMD3* gene analysis revealed no exonic mutations, which suggests genetic heterogeneity for BOS. Mechanisms for this heterogeneity include mutations in intronic sequences, promoter region defects, complete exonic deletion, or an entirely different gene defect.

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Anti–Bullous Pemphigoid 180 and 230 Antibodies in a Sample of Unaffected Subjects

Bullous pemphigoid (BP) is an acquired autoimmune bullous disorder characterized by circulating autoantibodies directed against basement membrane zone proteins BP180 and BP230. Serum BP180 autoantibody levels parallel disease activity. Although BP generally affects elderly patients, Wieland et al determined that the prevalence of circulating autoantibodies to BP230 and BP180 did not significantly increase with age in this cross-sectional study. These data suggest the possibility of a multifactorial pathogenic model for BP that includes triggers for disease initiation in susceptible patients.

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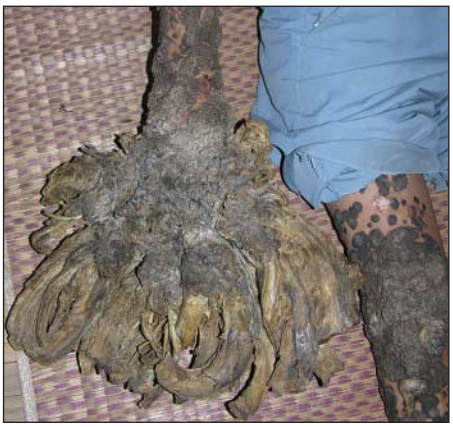
Disfiguring Generalized Verrucosis in an Indonesian Man With Idiopathic CD4 Lymphopenia



Human papilloma virus (HPV) infections cause a spectrum of clinical disease depending on the HPV strain as well as the host's immune status. Generalized verrucosis can be observed in patients with underlying immunodeficiency states that are acquired or congenital. In this case report, Alisjahbana et al describe an Indonesian man who was in good health until age 15 years, when a warty lesion appeared on his knee. The disfiguring lesions spread over much of his skin surface, encasing his hands, and preventing him from working. Laboratory analysis revealed HPV-2 infection and a severe immunodeficiency syndrome characterized by chronic $CD4^+$ T lymphocytopenia. These data suggest that patients presenting with massive cutaneous horns should be evaluated for HPV-2 infections and underlying immunodeficiency.

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Recent Trends in Systemic Psoriasis Treatment Costs

Psoriasis is a chronic autoimmune condition affecting up to 3% of the population. Therapeutic options vary, and one-third of Americans with psoriasis have moderate to severe disease that cannot be controlled with topical therapies alone. Systemic therapeutic options include 5 biological agents that are currently approved by the US Food and Drug Administration but are very expensive compared with conventional systemic agents. In this cost model study, Beyer and Wolverton demonstrate that systemic psoriasis therapy costs are increasing at a much higher rate than inflation, in part because of the trends in development of novel therapeutic agents for this disease.

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Incidence of Dermatomyositis and Clinically Amyopathic Dermatomyositis

Amyopathic dermatomyositis (DM) is a rare skin condition characterized by cutaneous symptoms of classic DM but lacking myopathy. Multiple studies have reported the incidence of cancer in patients with classic DM, but the incidence of classic or amyopathic DM remains largely unknown. In this retrospective population-based study, Bendewald et al demonstrate the overall incidence of DM to be 9.63 per million patients, with 21% of these cases representing the amyopathic subtype. Although the malignancy rate associated with DM was similar to that in previous reports, larger population-based studies are needed to estimate the malignancy risk with DM subtypes.

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