Overgrowth Syndromes

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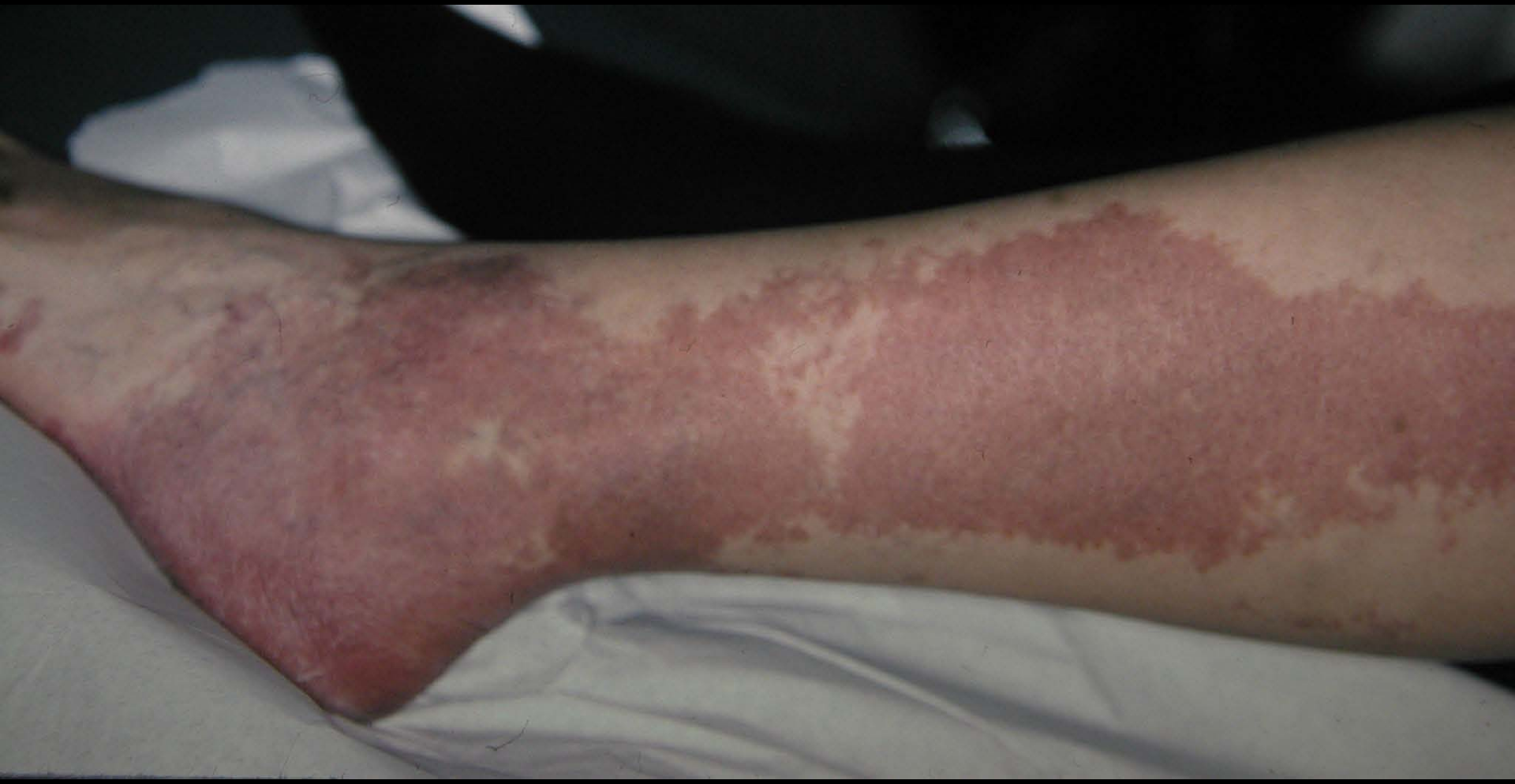
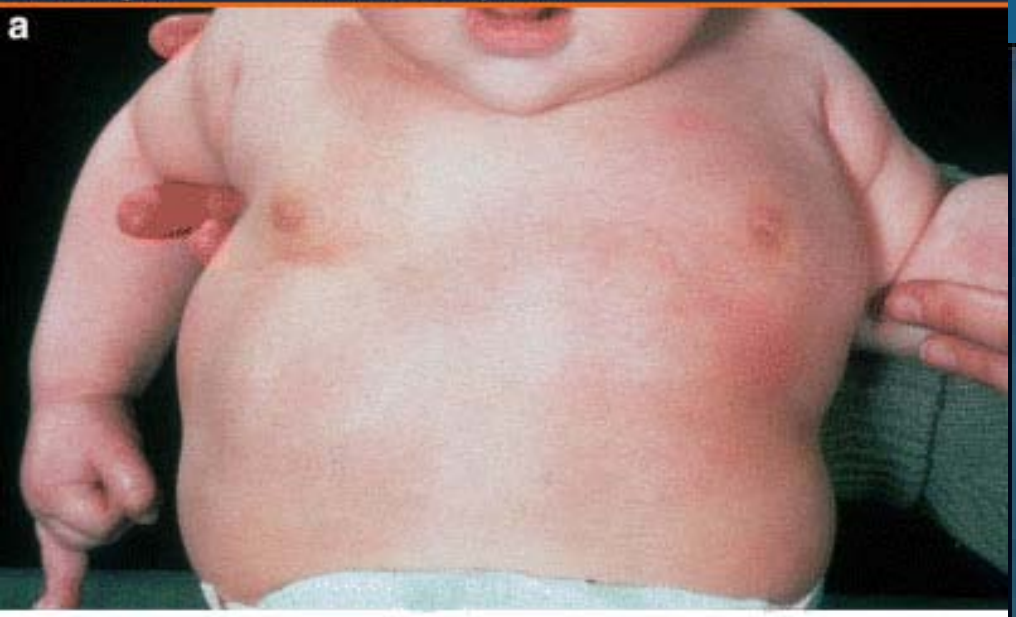
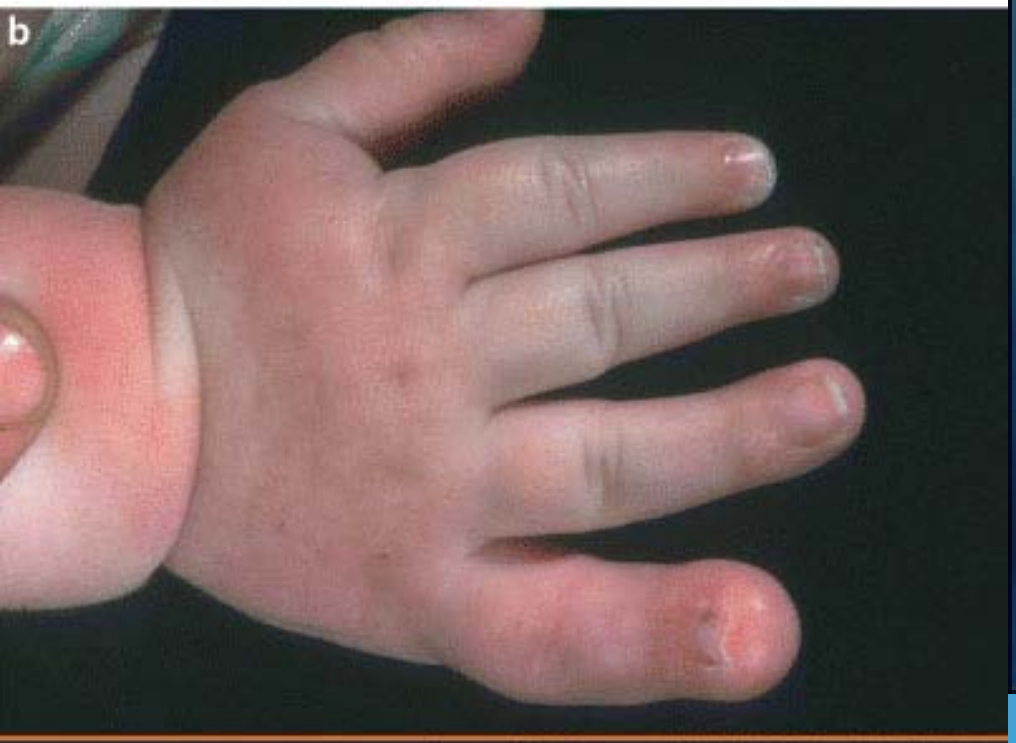


Klippel-Trenaunay Syndrome

Lesion	Appearance	Location	Onset	Pathology
Port wine stain	Irregular, macular, red to violaceous	Unilateral, lateral aspect of limb	Congenital, prenatal	Ectatic capillaries without endothelial proliferation
Varicose veins or venous malformations	Large, usually lateral superficial veins	Lateral aspect of limb, usually lower extremities, pelvic area	Congenital	
Lymphangiomas	Hyperkera-totic, verrucous frog-spawn like plaques	Usually superficial but can involve internal organs	Variable	





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Proteus Syndrome

Lesion	Appearance	Location	Onset	Pathology
Cerebriform connective tissue nevi	Gyriform, flesh-colored plaques	Palms and soles	Postnatal	Irregular collagenized fibrous tissue
Epidermal nevi	Tan or brown, flat topped hyperkeratotic to verrucous papules	Linear, follow Blaschko lines, typically on the trunk and neck	Congenital, prenatal	Acantholysis, hyperkeratosis, papillomatosis hyperplasia
Lipomas	Well demarcated, soft flesh-colored nodules	Abdomen, head, groin, legs	Postnatal	

Proteus Syndrome

Lesion	Appearance	Location	Onset	Pathology
Lipohypoplasia	Regions of skin with minimal fat, depressed violaceous plaques	Trunk, lower extremities	Postnatal	Loss of subcutaneous adipose tissue
Café au lait	Uniformly light or dark brown, sharply demarcated	Trunk, extremities	Variable	
Hyperpigmen-tation	Tan or brown, irregular macules	Linear, follow Blaschko lines	Variable	Increased melanin without increased melanocytes
Hypopigmen-tation	Hypopigmented macules	Face, trunk	Variable	Reduced melanin
Vascular malformations		Trunk, lower extremities	Congenital	

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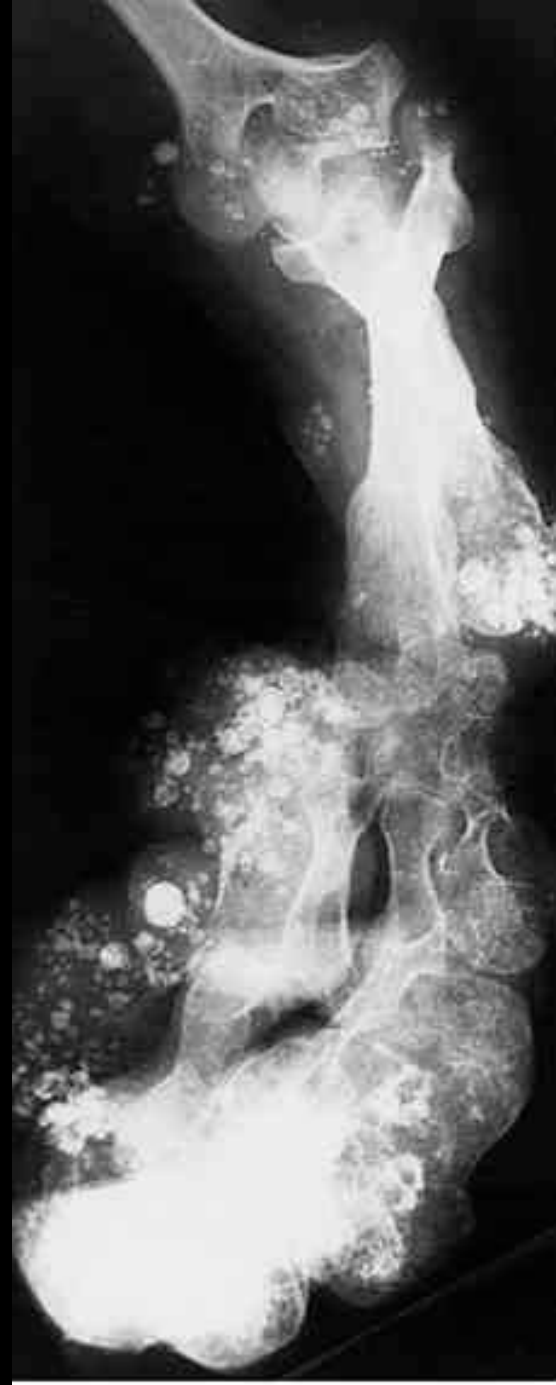

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Maffucci Syndrome

Lesion	Appearance	Location	Onset	Pathology
Hemangiomas	Blue compressible subQ nodules, do not completely blanch	Unilateral or bilateral	Birth to 30 years	Endothelial cell proliferation
Enchondromas	Irregular hard nodules	All parts of skeleton	Birth to 30 years	Lobules of intramedullary benign chondrocytes

References

- Gober-Wilcox JK, etal. Cutis 2009;83:255-262