
APRIL IOTADERMA (#302)

The words bead and beaded are found in dermatology in several instances. Can you remember three of them?

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Answer will appear in the *JAAD* Online section of the May issue of the Journal.

MARCH IOTADERMA (#301)

Question: Spice names are sometimes used in the descriptions of dermatologic diseases. For example, **cayenne pepper** is found in Schamberg's disease, **nutmeg** grater in lichen amyloidosis, and **salt** and **pepper** in scleroderma. Where would a dermatologist find "**cloves**" used?

Answer: Cloves is found in the acronym applied to an exceedingly rare, but distinctive, mosaic overgrowth syndrome with many dermatologic manifestations. **CLOVES** derives from the full name **C**ongenital **L**ipomatous **O**vergrowth-**V**ascular malformation-**E**pidermal nevi-**S**pinal anomaly syndrome. Dermatologists might encounter it when a patient presents with bizarre limb enlargements, or unusually distributed fatty tumors or large and deep vascular malformations, or large epidermal nevi masses. The cause of CLOVES syndrome has been found to be due to somatic [i.e. post zygotic] mutations in the PIK3CA gene, and thus various combinations of abnormalities can occur.¹⁻³ PIK3CA is a protein involved in the centrally important PI3K/AKT/mTOR signaling pathway.¹⁻³ A number of mutations in other parts of this pathway have been shown to produce diverse "overgrowth" phenotypes (PROS) including the Proteus, Klippel Trénaunay, Sturge-Weber, and hemimegaencephaly syndromes among others.² Treatment with a novel PIK3CA inhibitor has recently been shown to dramatically reduce overgrowths in a number of CLOVES patients.⁴

REFERENCES

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