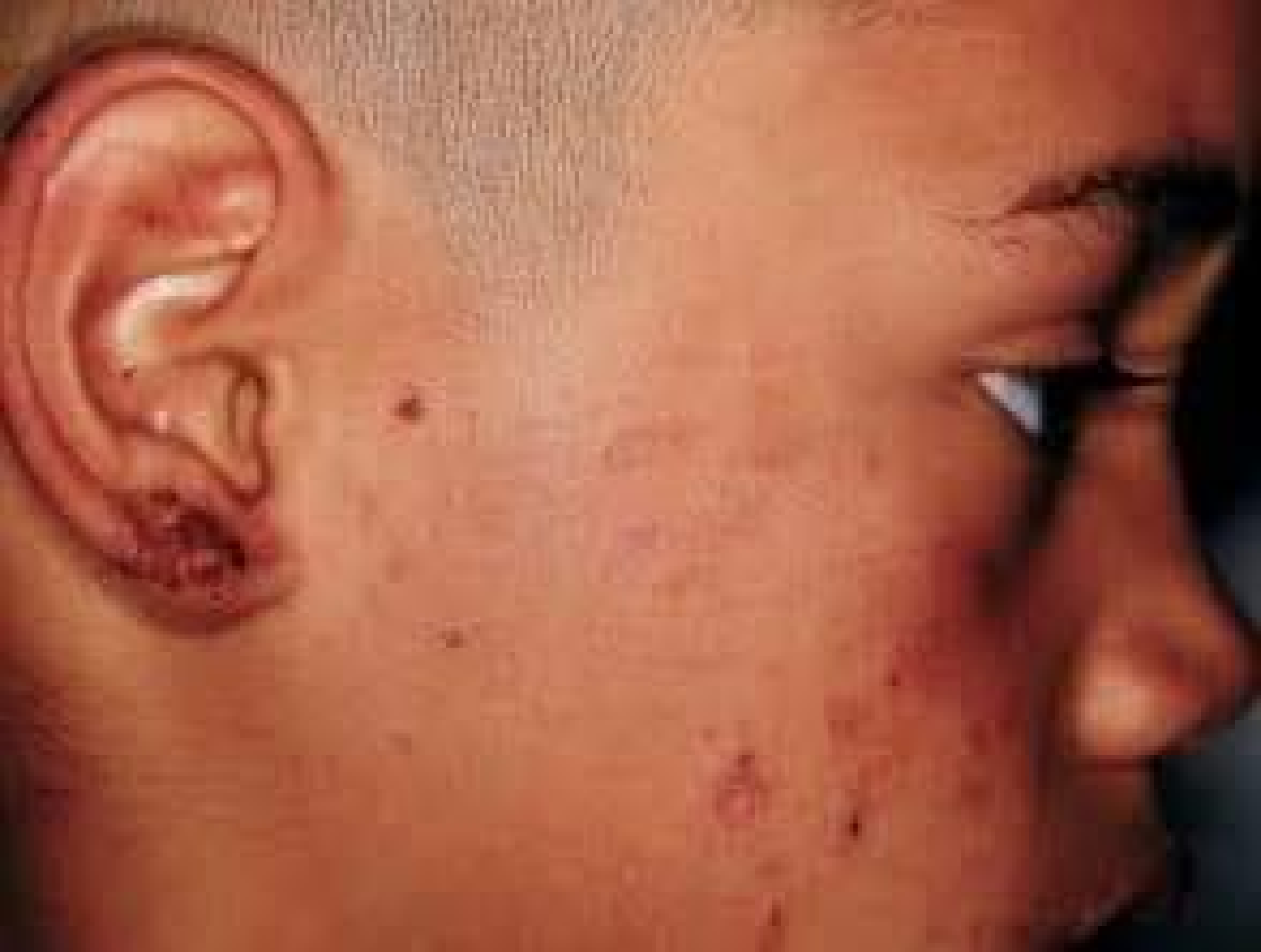
Hydroa Vacciniforme



HYDROA VACCINAFORME:

- Hydroa Vacciniforme is a very rare photosensitivity/auto-immune disorder.
- People who have HV don't have the ability to absorb UV rays. It's basically like an extremely severe allergy to the sun.
- When the skin is exposed to the sun, the skin develops fluid-filled blisters that quickly puff and scab, then heal with pox-like scars.
- As of now, there are around 300 people in the world that have been diagnosed with Hydroa Vaccinaforme.

HYDROA VACCINIFORME

- Hydroa vacciniforme (HV) is a very rare photodermatosis of unknown etiology that principally starts in childhood.
- characterized by recurrent crops of papulovesicles or vesicles most commonly on the face and the dorsa of the hands but other sun-exposed areas of the skin may also be involved. The vesicles resolve with pocklike scarring.

Hydroa Vacciniforme

- Photodermatosis with onset in childhood
- Lesions appear in crops with disease free intervals
- Attacks may be preceded by fever and malaise
- Ears, nose, cheeks, and extensor arms and hands are affected
- Within 6 hrs of exposure stinging may occur

- **Hydroa Vacciniforme (HV):**

❖ **C/F:**

- boy > girls, mean age 8 yrs, resole spont. in adulthood
- Recurrent crops of discrete 2-3mm erythematous macules evolve into blisters hrs to a 1-2 days after sun exposure.
- Healing occurs within days, with umbilication followed by crusting & pitted, varioliform scarring. Adverse effect on quality of life.
- Rare presentations ocular involvement (keratoconjunctivitis & uveitis) & blistering of the lips.

❖ **Histopathology:**

❖ **Phototesting:** UVA in monochromatic phototesting…papulovesicular

❖ **Pathophysiology:** not clarified. (Latent EBV infection), some cases reported in adult (fatal) different from that of children.

❖ **D.Dx:** EPP, vesicular PMLE, bullous SLE, & PCT.

❖ **Treatment:** Photoprotection, Chloroquine, beta-carotene & PUVA. In severe cases, systemic corticosteroid.