Huriez Syndrome

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- **History**
  - 31 year-old male
  - Rapidly growing painful tumor on right index finger
  - Undiagnosed chronic skin disorder involving his hands that began in his teens and had never been treated
  - Two episodes of meningitis in childhood

- **Family history**
  - No family members had similar problems
Huriez Syndrome

- Sclerotylosis
- Triad
  - Scleroatrophy
  - Hypoplastic nail changes
  - Palmoplantar keratoderma
- No history of Raynaud’s phenomenon
- Hypohidrosis
- Increased risk of SCCs
**Squamous cell carcinomas**

- Increased risk of SCC’s
- Age 30-50
- Increased mortality rate from metastatic squamous cell carcinoma (5%)
- Multiple numbers of SCC’s in individual patients
- Reduced number of Langerhans cells _ decreased immune surveillance?
- Normal excision-repair mechanism
- No clear internal malignancy risk

**Inheritance**

- Autosomal dominant
- First described in 1968 in two families from Northern France, since has only been reported in a handful other families
- 27/114 family members
- Scattered case reports
- Linkage analysis mapped the gene to chromosome 4q23
Treatment

- Close surveillance for malignancy
- Retinoids

Conclusions

- Rare genetic dermatosis
- Only palmoplantar keratoderma associated with cutaneous malignancy
- Appropriate screening and aggressive treatment
Thanks!

- Ben Ehst
- Alfie Krol
- Rodd Takiguchi

Questions?
References

- Huriez C, Deminatti M, Agache P, Mennecier M. Une g_nodysplasie non encore individualis_e: la g_nodermatose scl_ro-atrophiant e k_ratodermique des extremit_s fr_quemment d_g_nerative. Sem Hop 1968;44:481-8.