6 year-old girl, sub-cutaneous flesh-coloured smooth nodules on the right fingers

Presumptive diagnoses:
- Infantile myofibromatosis?
- Rheumatoid nodules?
- Granuloma annulare?
What is your diagnosis?
Careful follow-up (x-ray)

- **New lesions** developed on the hand, the ankle and the elbow
- At the age of 12, occurrence of multiple enchondromas

↓

Maffucci’s syndrome

Bodemer C, Fraitag S, Amoric JC et al:
Spindle cell hemangioma

- Has been previously described as «spindle cell hemangioendothelioma» since the initial cases recurred many times.
- It is now considered as a benign disorder.
- It remains controversial.
  - Vascular neoplasm
  - Malformative process with venous thromboses
- Can be isolated but can be the sentinel event for Maffucci’s syndrome.
Maffucci’s syndrome

- Non hereditary disorder
- Diffuse asymmetric enchondromatosis associated with
  - multiple venous malformations
  - various musculoskeletal deformities
- Cutaneous and mucosal venous malformations appear in early infancy ➔ deformations
- Risk of malignant transformation of the enchondromas into chondrosarcomas (15 %)
- And other malignant neoplasms
  - Fibrosarcomas
  - Angiosarcomas
  - Osteosarcomas
  - Malignant ovarian neoplasms…

⇒ Follow-up is mandatory!
### Vascular Tumors

- Infantile hemangioma
- Congenital hemangiomas (NICH and RICH)
- Tufted angioma
- Kaposiform hemangioendothelioma
- **Spindle cell hemangioma**
- Others, rare (Dabskas’s T..)

### Vascular Malformations

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### Complex-combined vascular malformations

- CVM, CLM, LVM, CLVM, AVM-LM, CM-AVM