UNIVERSITÄTS-
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Interesting cases – lessons learnt

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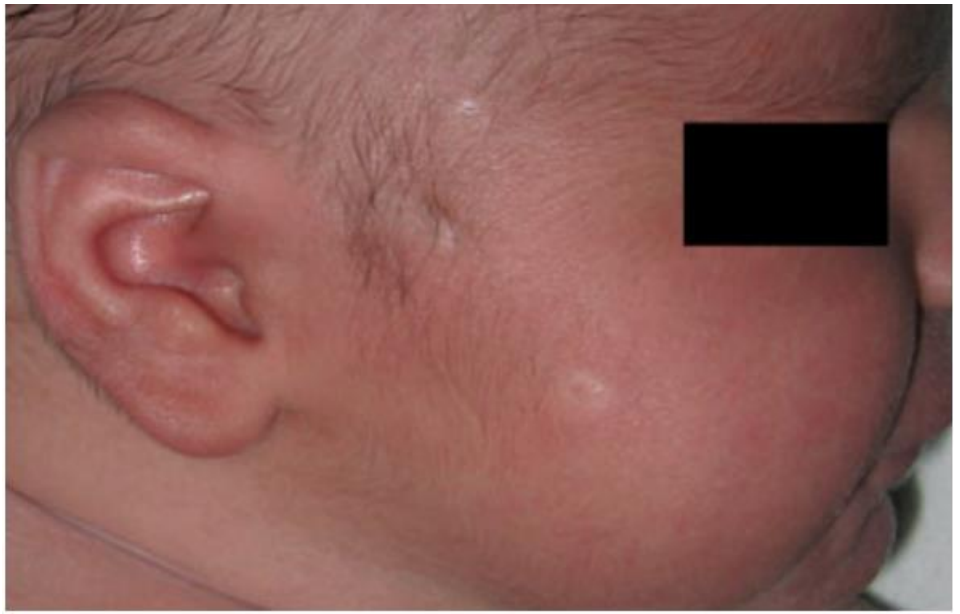
Case 1

Referred for the evaluation of **bilateral hypopigmented lesions** on the face present since birth

- Female
- 1st child, family history unremarkable
- Uneventful pregnancy, born full term
- Postnatal period normal with age-appropriate development
- At the age of 2 months diagnosis of SVT – stable under propranolol therapy



Krathen et al, ped.Dermatol 2008

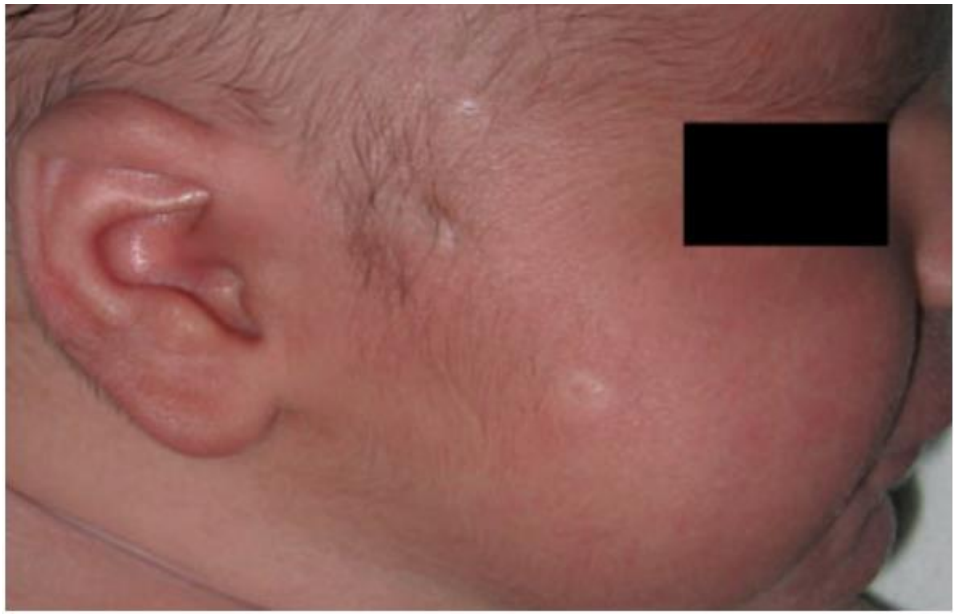
First consultation
Age 4 month

Complete physical examination:

- *Circular hypopigmented atrophic macules linearly arranged on both cheeks*

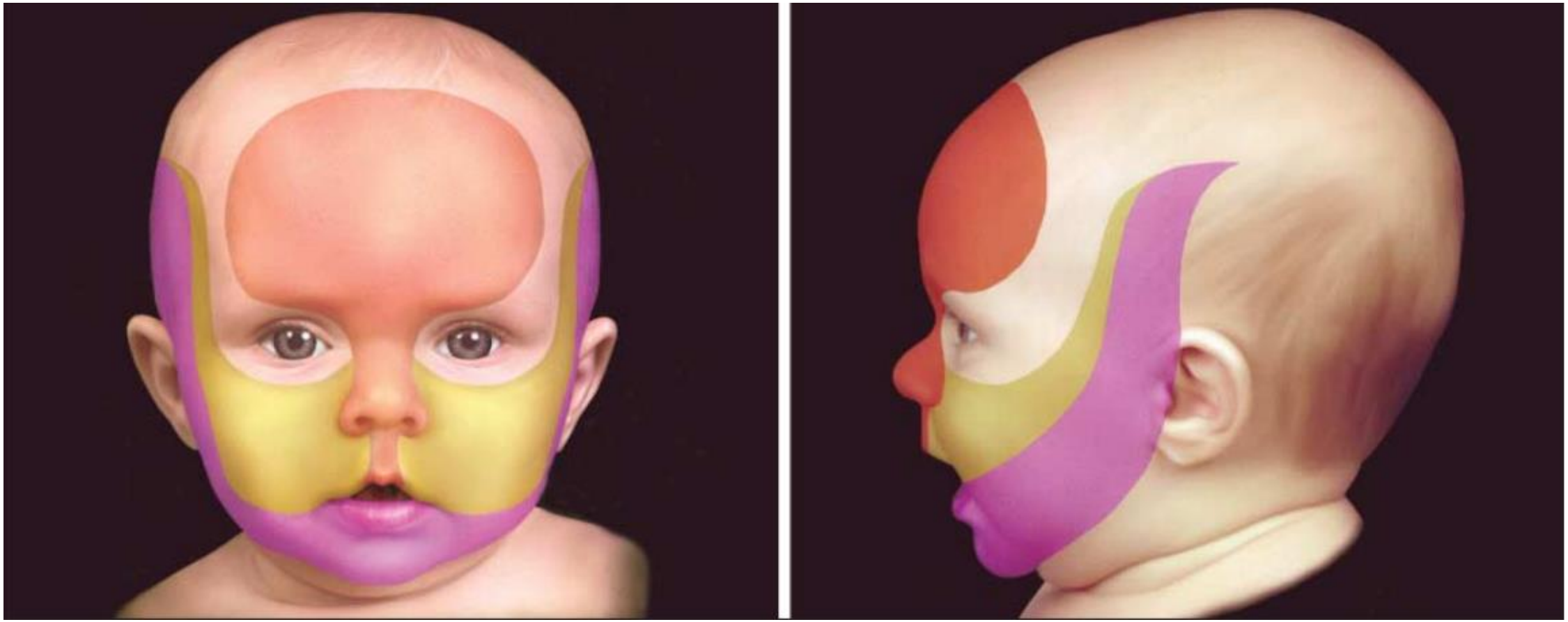
- No other abnormalities of the skin, hair or nails

- No dysmorphic signs



Krathen et al, ped.Dermatol 2008

Facial embryonic fusion lines



Waner et al, Arch Dermatol 2003

Focal preauricular dermal dysplasia (FPDD)
Focal facial dermal dysplasia (FFDD)

- FPDD is a form of aplasia cutis congenita in which bilateral preauricular atrophic skin lesions occur caused by defective fusion at the **maxillary and mandibular prominences**

- FFDDs are a group of rare inherited developmental disorders chararacterized by congenital scar-like lesions in the **bitemporal (FFDD1, 2 and 3)** or **preauricular (FFDD4)** areas.

- **FPDD (FFDD4)** is an autosomal recessive trait without dysmorphic findings or extracutaneous anomalies ***(CYP26C1-mutation)***

- Biopsy not necessary for diagnosis, treatment options limited (surgical excision)

Krathen et al, Pediatr Dermatol. 2008
Slavotinek et al, Hum Mol Genet 2012

Case 2

Referred for the evaluation of a **periumbilical abdominal ash** that was noted at age of 2 weeks

- Female
- Born full term
- Uneventful pregnancy, healthy parents
- Postnatal period normal with age-appropriate development

Complete physical examination:

- Afebrile girl with an umbilical stump surrounded by a
- ***Reticular blanchable erythema with teleangiectasias on the bilateral abdomen***
- No evidence of infection in or surrounding the umbilicus

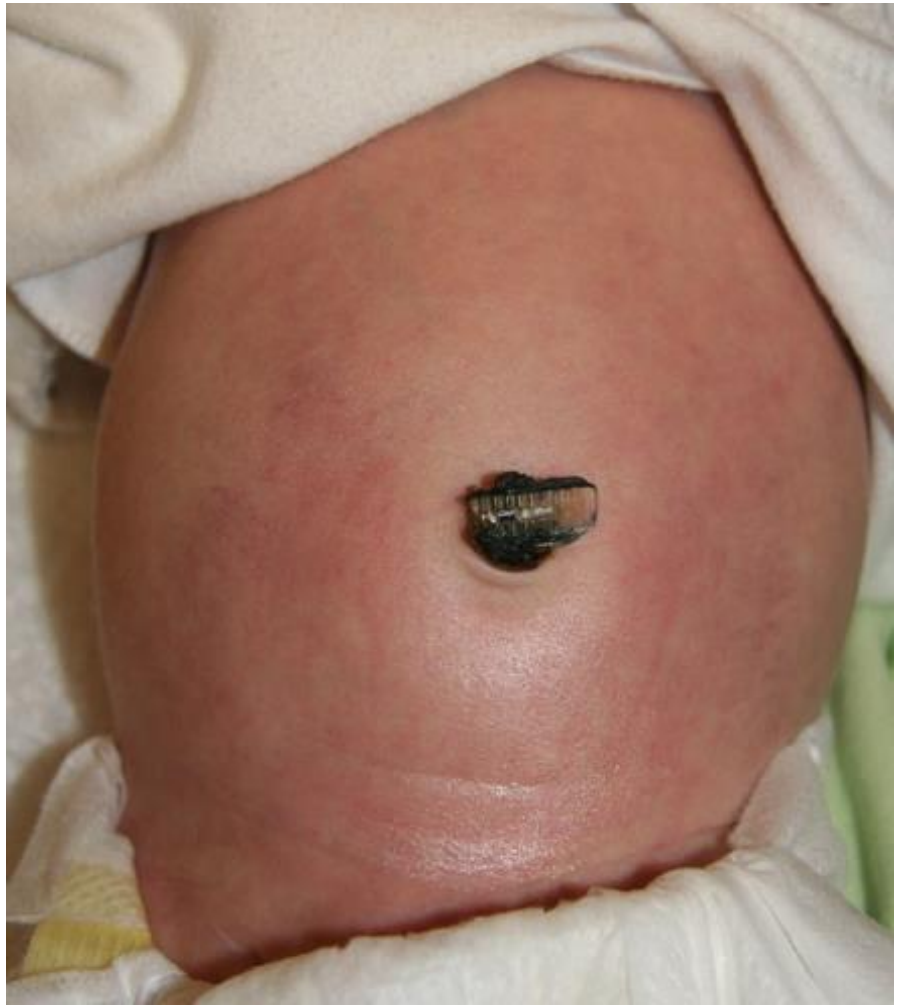
What is your differential diagnosis and what do you do next?

a. You think about an Erythema toxicum neonatorum – and explain its harmlessness to the parents
b. You fear an omphalitis – and send the family to the emergency
c. You diagnose a capillary malformation – no further examination

Self-limited neonatal periumbilical erythema

- Benign self-limited condition
- Periumbilical reticular blanchable erythema
- Inflammatory and vasodilatory response during the normal umbilical cord separation (exact pathophysiology not known – cytokine release)

- No clinical sign of intra- or periumbilical infection (fever, umbilical discharge, abdominal tenderness)
  - ***CAVE: Omphalitis***

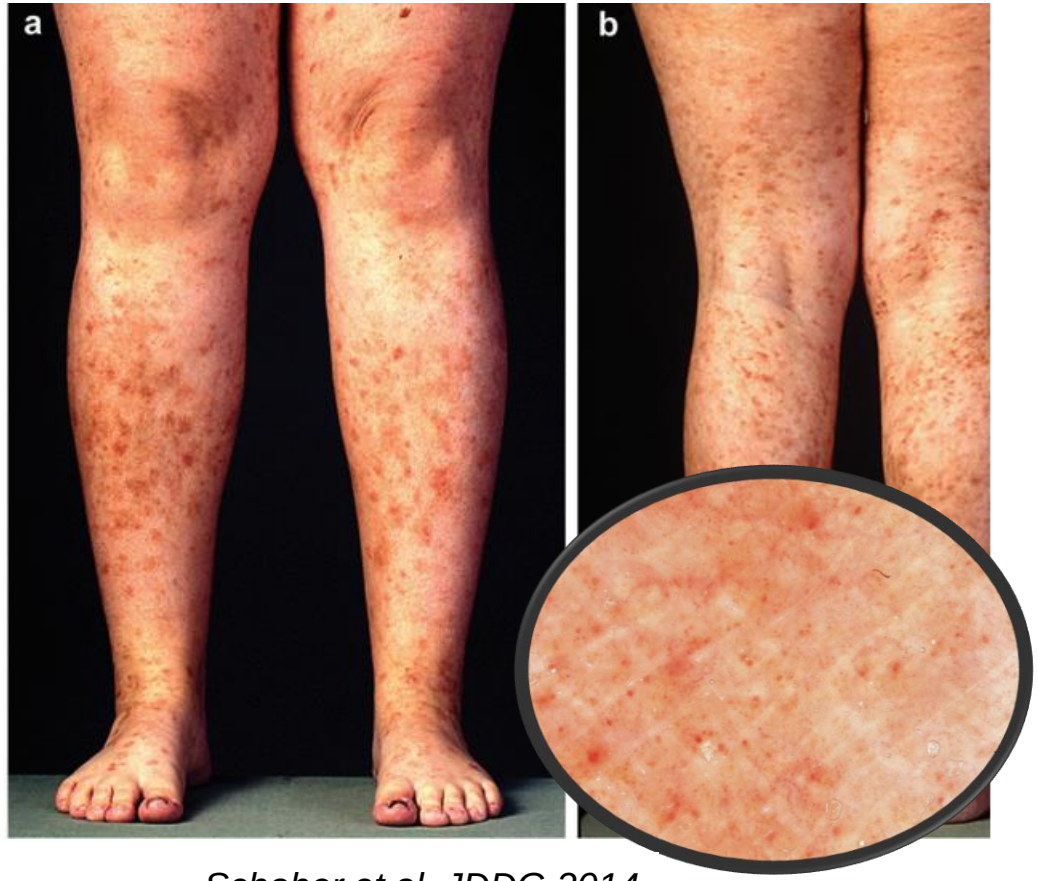


Peterman et al, Pediatr Dermatol. 2017

Case 3

Referred for the evaluation of **red-brown reticular macules** distributed on the lower limbs and on the abdomen first appeared at the age of 9 months

– Male, born full term
– Uneventful pregnancy, healthy parents
– Postnatal period normal with age-appropriate development



Schober et al, JDDG 2014

Complete physical examination:

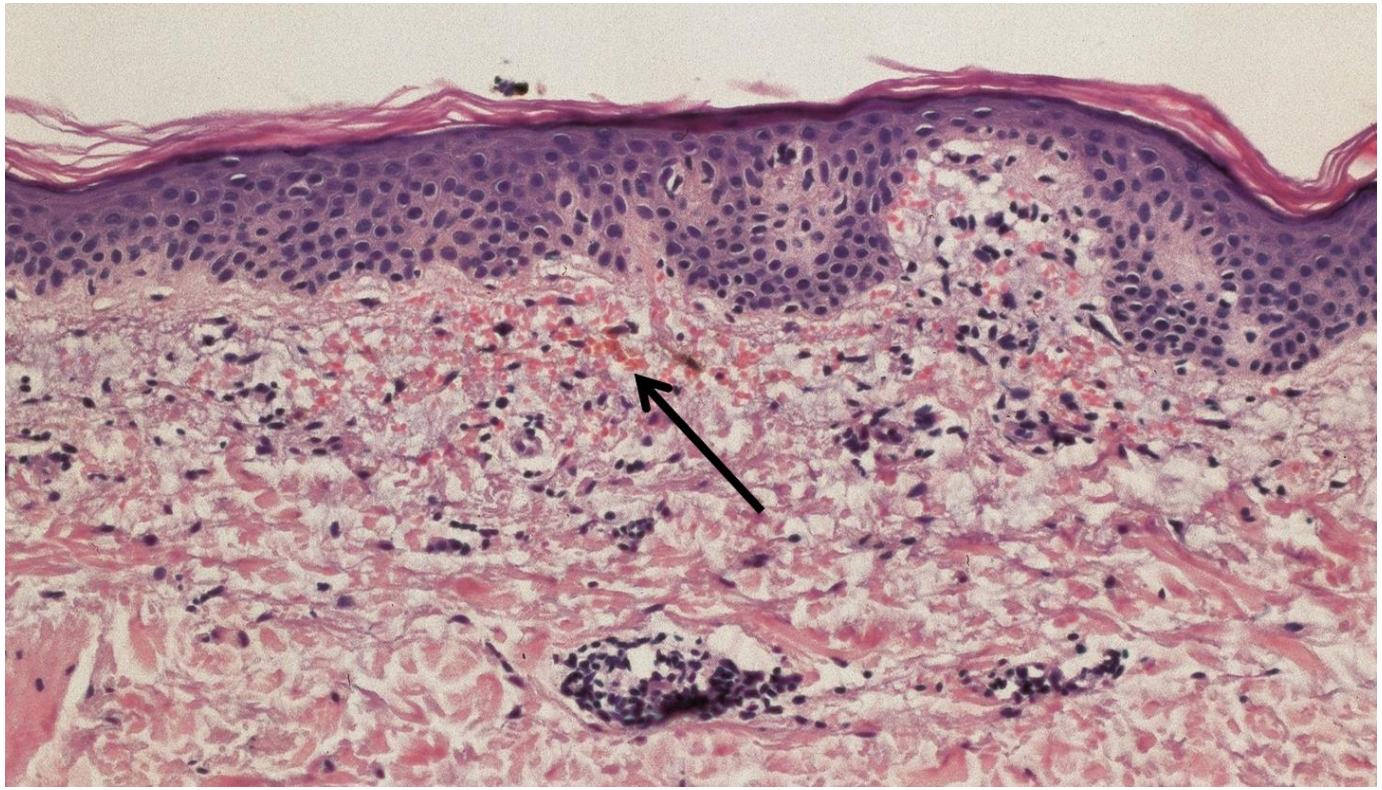
– well-appearing afebrile boy
– ***Purpuric erythematous macular eruption on the lower extremities and abdomen***
– asymptomatic, no history of infection in the last weeks

What is your differential diagnosis?

a. Small vessel vasculitis
b. Coagulopathy
c. Vascular malformation
d. Pigmented purpuric dermatosis

Further investigations

- **Full blood count and coagulation**
  - Normal, no thrombocytopenia
- **Biopsy- Histology**
  - Lymphocytic perivascular infiltrate in the dermis with extravasation of
  - Erythrocytes and hemosiderin deposition
  - **No fibrinoid necrosis of the vessels**



Pigmented Purpuric Dermatosis (PPD) – M. Schamberg

- Proposed pathophysiology – lymphocytic capillaritis
- Reddish-brown macules bordered by punctate „cayenne pepper“ spots
- Primarily a clinical diagnosis but biopsy recommended to rule out vascultits and CTLC

- Lichen aureus and M. Schamberg disease most common subtypes of PPD in children
- Usually healthy children, most often no clear trigger, parainfectious?
- M. Schamberg often exanthemateous
- Asymptomatic (seldom pruritus, cosmetic concerns), **no correlation with systemic disease**

Torrelo et al, J Am Acad Dermatol 2003

Schober et al, JDDG 2014

PPD in children – clinical course and treatment options

- PPD is a benign disorder with high rates of complete resolution (75% within 1 year)
- Chronic recurrent course possible
- There are currently no well-established treatments for PPD
  - Topical and systemic corticosteroids, photothterapy (UVBnb)
  - **Rutoside in combination with Vitamin C (antioxidative radicals)**

Original Article

Pigmented purpuric dermatosis in children: a retrospective cohort with emphasis on treatment and outcomes

A. Ollech ✉, A.S. Paller, L. Kruse, B. Kenner-Bell, S. Chamlin, A. Wagner, L. Shen, R. Yousif, L.C. Balmert, A.J. Mancini

However, no advantage when compared to watchful waiting

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Ollech et al, JEADV 2020

Schober et al, JDDG 2014

Case 4

Referred for the evaluation of a **erythematous violaceous solitary** plaque on the abdomen present since birth

– Healthy girl, born full term, family history unremarkable

– Postnatal period normal with age-appropriate development

Complete physical examination:

– Well-appearing girl

– ***Erythematous-violaceous solitary slight indurated plaque on abdomen (1.5x3.5cm)***

– No growth, no thrill, no other vascular stains

Punchbiopsy – Tufted angiom

ISSVA classification for vascular anomalies ©

(Approved at the 20th ISSVA Workshop, Melbourne, April 2014, last revision May 2018)

Vascular tumors

Benign

- Infantile hemangioma
- Congenital hemangioma
- Pyogenic granuloma
- **Tufted angioma**

Locally aggressive or borderline

- Kapos. hemangioendothelioma

Malignant

Vascular malformation

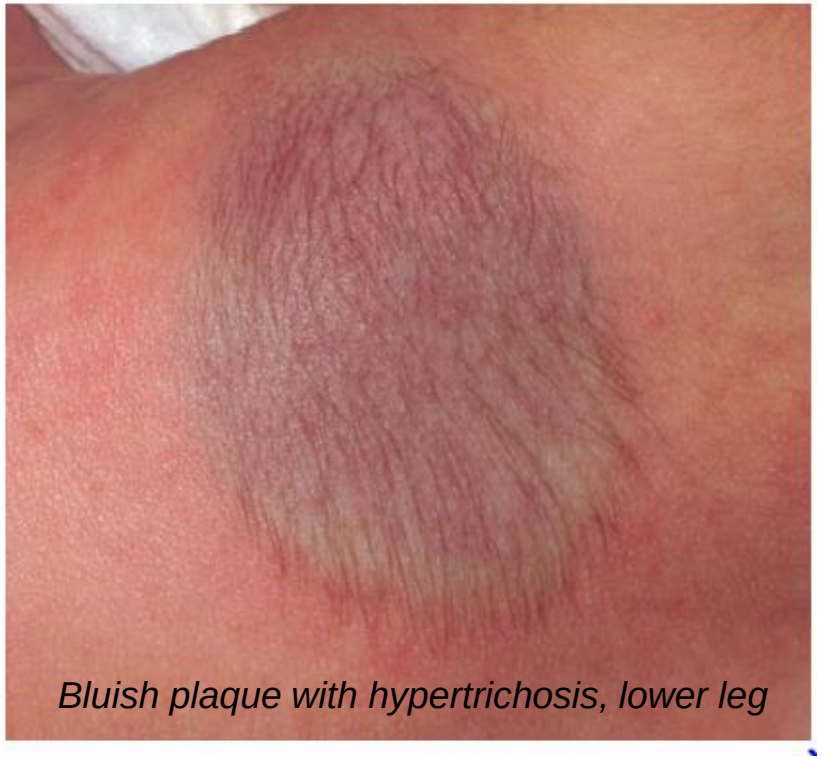
Simple

- Capillary malformations
- Lymphatic malformations
- Venous malformations
- Arteriovenous malformations

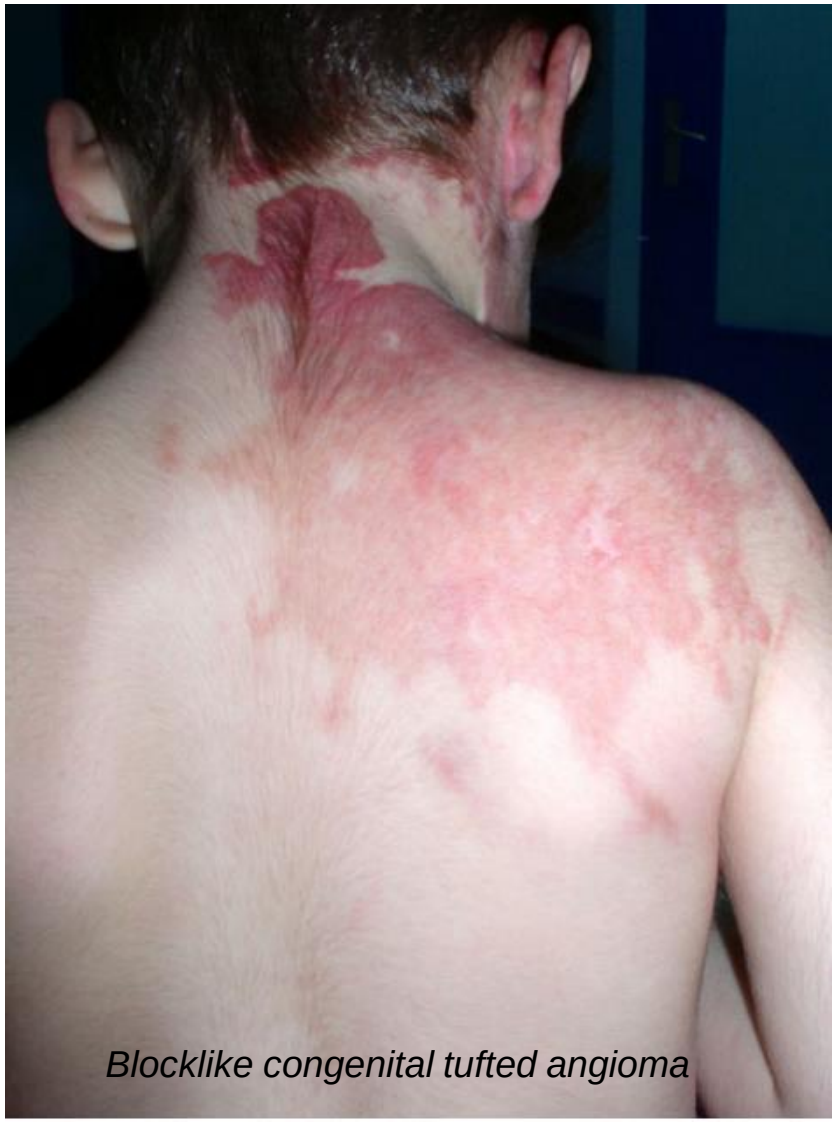
Combined

Associated with other anomalies

Tufted angioma- clinical spectrum



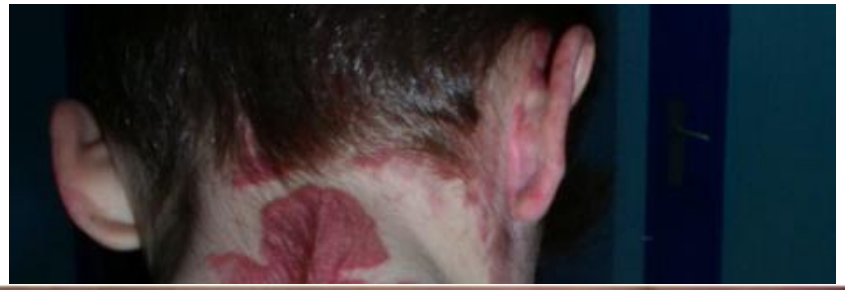
Bluish plaque with hypertrichosis, lower leg

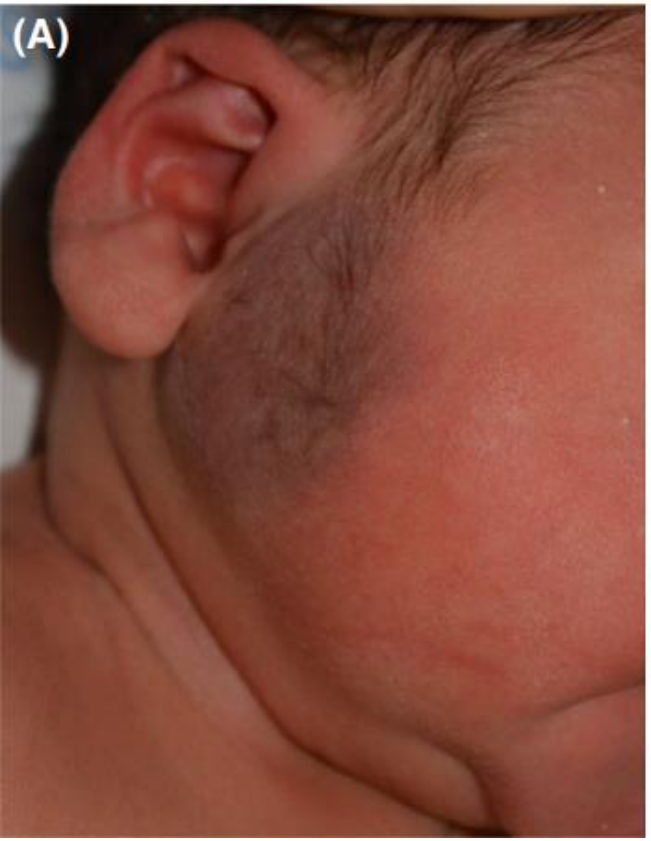


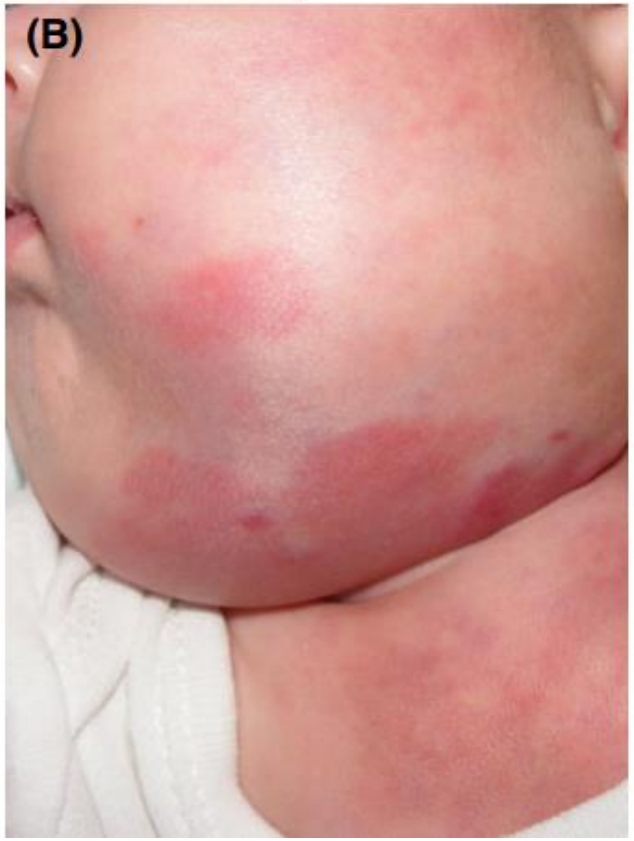
Blocklike congenital tufted angioma

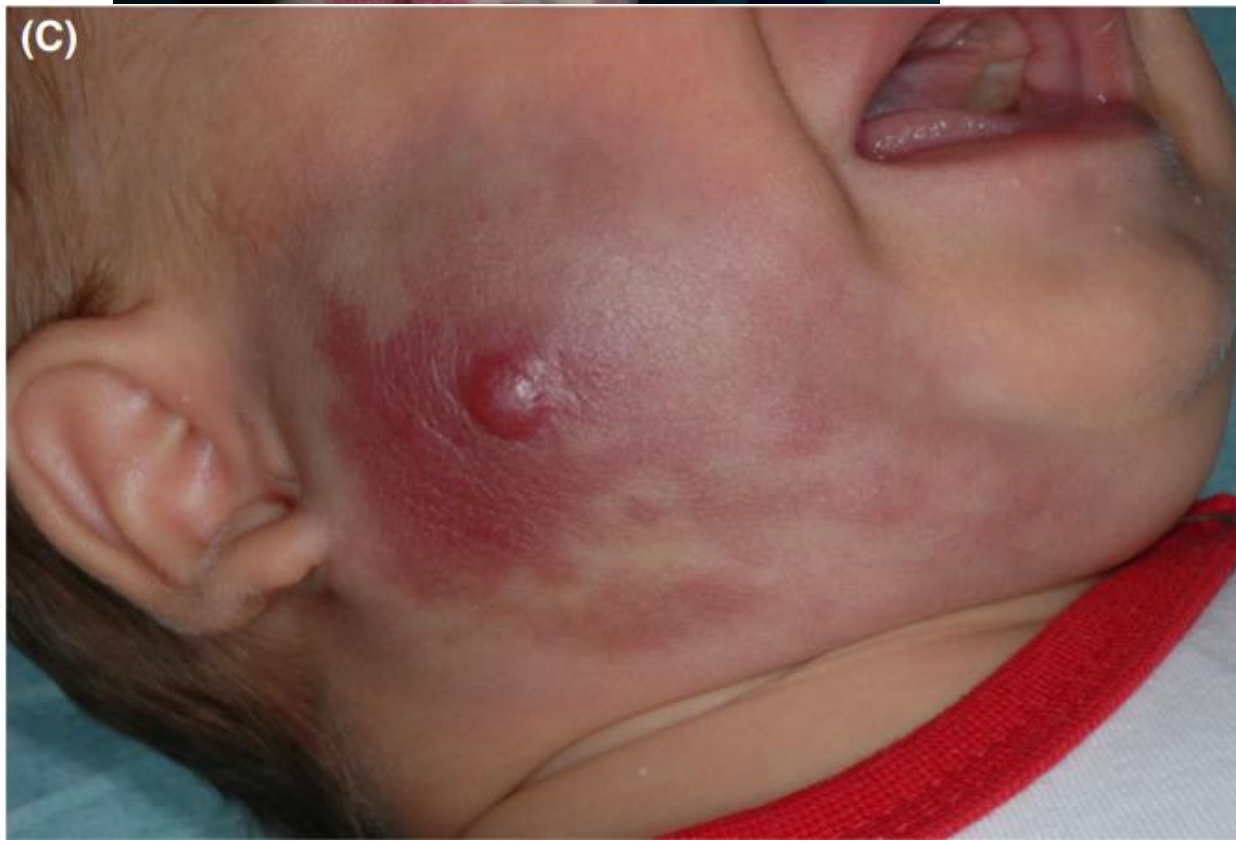
Feito-Rodriguez M et al, Pediatr Dermatol 2018

Tufted angioma- clinical spectrum









Feito-Rodriguez M et al, Pediatr Dermatol 2018

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- **Kapos. hemangioendothelioma**

Malignant

Vascular malformation

Simple

Kasabach-Meritt Phenomenon (KMP)

- profound thrombocytopenia
- raised D-dimers
- low fibrinogen

Associated with other anomalies

Further investigations

- **Ultrasound-Doppler of the lesion and abdomen**
  - normal

- **Haemato-Oncology consultation**
  - Differential blood count and coagulation
    - both normal
  - transient elevation of the D-Dimers

  →no need for further investigations

- Clinical follow-up 3-6 monthly (watch & wait strategy)
  - **Urgent assessment if clinical signs of bleeding (petechiae or mucosal bleeds)**