Inflammatory dermatoses

Case n°2

S Fraitag
Necker-Enfants Malades Hospital
Paris, France
- 33 YO woman
- No relevant medical history
- Cutaneous lesions extending gradually for 15 years on the lower extremities
Small, asymptomatic, violaceaus, slightly purpuric, bilateral and symmetrical, papules
Suggested clinical diagnosis: lichen planus
Diagnosis?

Progressive mucinous histiocytosis, sporadic form
Progressive mucinous histiocytosis

- Rare, benign, non-Langerhans cell Histiocytosis
- First described by Bork and Hoede in 1988, 21 cases have been reported
- Exclusively localized in the skin
- There are 2 forms:
  - hereditary
  - sporadic
Clinical characteristics

- 1- to 3-mm smooth, and often shiny papules. Lesions up to 10 mm in diameter have been reported.
- May be reddish-brown, reddish-yellow, pink, or skin-colored.
- Areas of predilection: the scalp, face, dorsal hands, forearms, thighs, and fingers.
- Lesions are non tender, mild pruritus has been described.

Acta Derm Venereol 2010; C. Schlegel et al.
British Journal of Dermatology 1999; D Wong et al.
Hereditary form

- 17 cases reported in a total of six families
- Affects mostly females (only 3 cases in males)
- An autosomal dominant inheritance is presumed
- Begins in childhood or adolescence and progresses slowly
Sporadic form

- Same presentation, later onset
- Distributed widely across four continents

<table>
<thead>
<tr>
<th>Sex</th>
<th>Site</th>
<th>Age at onset</th>
<th>Références</th>
</tr>
</thead>
<tbody>
<tr>
<td>F</td>
<td>Face</td>
<td>41 YO</td>
<td>A. Young et al, J Cutan Pathol 2006, Baltimore</td>
</tr>
<tr>
<td>F</td>
<td>Knees</td>
<td>50 YO</td>
<td>A. Young et al, J Cutan Pathol 2006, Baltimore</td>
</tr>
<tr>
<td>F</td>
<td>Dorsum of the hands</td>
<td>adolescence</td>
<td>U. Sass and al, Br J Dermatol 2000, Brussels</td>
</tr>
<tr>
<td>F</td>
<td>Head and forearms</td>
<td>28 YO</td>
<td>Narváez-Rosales and al, Skinmed 2013, Mexico</td>
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</tbody>
</table>
Histopathology

- Stereotyped
- Well circumscribed lesion in the upper and mid dermis
- Non affected epidermis
- Accumulation of histiocytes accompanied by some mast cells
- Embedded in an abundant mucinous deposit
## Immunohistochemistry

<table>
<thead>
<tr>
<th>Our case</th>
<th>Published cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>CD68 +</td>
<td>CD68 + or -</td>
</tr>
<tr>
<td>FXIIIa +</td>
<td>FXIIIa + or -</td>
</tr>
<tr>
<td>CD163 +</td>
<td>CD163 not done</td>
</tr>
<tr>
<td>CD1a -</td>
<td>CD1a -</td>
</tr>
<tr>
<td>PS100 -</td>
<td>PS100 -</td>
</tr>
</tbody>
</table>

Non Langerhans-cell Histiocytosis
Non-Langerhans Cell Histiocytoses

- Cutaneous non-LCH
  - Xanthogranuloma family
    - Juvenile xanthogranuloma
    - Adult xanthogranuloma
    - Solitary reticulohistiocytoma
    - Benign cephalic histiocytosis
    - Generalised eruptive histiocytosis
    - Progressive nodular histiocytosis
    - Progressive mucinous histiocytosis
  - Non-xanthogranuloma family
    - Solitary cutaneous Rosai-Dorfman-Destombes disease

- Cutaneous non-LCH with a major systemic component
  - Xanthogranuloma family
    - Xanthoma disseminatum
  - Non-xanthogranuloma family
    - Multicentric reticulohistiocytosis
    - Necrobiotic xanthogranuloma
Non-Langerhans Cell Histiocytoses

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    - Necrobiotic xanthogranuloma

Dermal dendrocyte

FXIIIa +

Weitzmann  Pediatr Blood Cancer 2005
Zelger B Br J Dermatol 2001
Electron microscopy

Numerous circular intra-cytoplasmic myelin bodies and zebra bodies

Closely resembling a lysosomal storage disease

Pathogenesis

- Unknown until now

- Storage disease hypothesis:
  - Hereditary cases
  - Progressive evolution
  - Myelin bodies in EM

- But in described cases:
  - Lipid metabolism normal
  - Lysosomal enzyme dosage normal
Clinical differential diagnoses

- Lichen planus
  - Itchy

- Angiohistiocytoma with multinucleated cells:
  - Women over 50 YO

- Other non langerhans-cell histiocytoses
  - Generalized eruptive histiocytosis: spontaneous recovery after some years
Mainly mucinoses:
- **Acral papulous mucinosis:**
  - Mainly women, outbreaks of multiple flesh-colour 2 to 5 mm papules, symmetrical, on the dorsum of the hands and wrists extending progressively
  - Large pools of mucin deposits on the superficial and mid dermis
  - **Fibroblastic cells but no histiocytic accumulation**
Outcome

- Progressive multiplication of the lesions
- Extending to other areas of the body
- Remains benign, only restricted to skin
- Esthetic issues
- No efficient treatment (laser ?)

Think of progressive mucinous histiocytes: histiocyte accumulation + mucin deposition!
Some beautiful Paris bridges

Le pont Alexandre

Le pont neuf

Le pont des Arts

Le pont Mirabeau

Le pont Charles de Gaulle

Le pont Louis-Philippe