A rare cutaneous condition - a case presentation

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A 12 year old African male patient presented with asymptomatic multiple skin lesions arising on his (L) buttock – duration of 1 year. The mother is of the impression that the lesions are progressive in nature since the onset. No relevant past or family history was elicited. No history of trauma given. Following my clinical examination, a 8mm punch biopsy was carried out under LA, after consent was taken from the parents.
The histopathology was reported as –

- `focal attenuation of the epithelium overlying masses of fat in the upper dermis`

**Histopathological Diagnosis**

- Features suggest an *Intradermal Lipoma*
HISTOLOGY OF NORMAL SKIN

- Thick Skin
- Epidermis
- Dermis
- Hypodermis
Figure 2: Photomicrograph showing ectopic fat (arrow) in the mid and lower dermis (H/E, 40x)
Lesions
Lesions
What is this condition?
Lipomas by definition are the most common neoplasms of mesenchyme. Common lipomas consists of mature fat cells throughout. Most lipomas (98%) occur as single or multiple subcutaneous growths.

They come to clinical attention only if they reach inordinate size or are perceived as blemishes in need of cosmetic removal.
1. Hamartomas
   - Nevus lipomatosus cutaneous superficialis
   - Folded skin with lipomatous nevus
   - Congenital lipomatosis - Proteus syndrome

2. Benign Neoplasms of white fat
   - Homogenous - Mature Lipoma
3 - With heterogeneous mesenchymal elements:

1. Fibrolipoma
2. Myxolipoma
3. Ossifying lipoma
4. Multiple lipoma syndromes
5. Adiposis dolorosa
6. Benign symmetric lipomatosis
7. Angiomyolipoma
4 - With **specific clincopathological** settings:

1. Perinueral lipomas
2. Lipoma of tendonsheath & J oints
3. M ultiple lipoma syndromes
4. Adiposis dolorosa (Dercums disease)
5. Benign symmetric lipomatosis
6. Familial multiple lipomatosis
Continued

- 5 - Pseudosarcomatous benign adipose tissues
  1. Spindle Cell Lipoma
  2. Pleomorphic lipoma
  3. Chondroid Lipoma
  4. Lipoblastoma & Lipoblastomatosis
  5. Inter- & Intramuscular Lipomas
  6. Cervical symmetrical lipomatosis
  7. Pelvic lipomatosis
  8. Diffuse Lipomatosis

6 - Benign Neoplasms of brown fat
- Hibernoma
Taking into account the age of the patient, onset of the lesions, the clinical appearance & anatomical location of the lesions and the histopathological report.....a diagnosis of **NEVUS LIPOMATOSUS CUTANEOUS SUPERFICIALIS OF HOFFMAN & ZURHELLE** appears the most consistent.
Nevus lipomatosus cutaneous superficialis

- (NLCS) is a rare idiopathic hamartomatous anomaly, the classic type of which presents with asymptomatic grouped, soft, skin-colored to yellow papules and nodules within the first three decades of life.

- (HAMARTOMA - tumor-like collection of excess tissue or abnormally situated tissue formed during development)

- There is no familial tendency, nor sex predilection.

- Histology is characteristic with ectopic mature adipose tissue within the dermis.
Clinically there are two types. The classic (or multiple) type usually presents within the first 3 decades of life with clusters of soft, fleshy skin colored or yellow nodules having either smooth and wrinkled or cribiform and peau d'orange appearance, located most commonly on the lower trunk, especially on the back, buttocks or hips or abdomen, and on the upper posterior thighs.
Rare involvement of the face or scalp have been reported. They are generally present at birth, but may first appear during childhood or adolescence.

They are almost invariably asymptomatic, although occasionally ulceration may occur.

The solitary form of NLCS usually appears during the third to sixth decades of life as a single papule or nodule without a specific location.

There are reports of coexisting café-au-lait macules, leukodermic spots, over lying hypertrichosis, and comedo-like alteration.
A recent report described a giant NLCS with multiple folliculosebaceous cystic hamartomas and dermoid cysts. A case of NLCS with localized scleroderma like appearance has been reported.

The differential diagnosis before biopsy may include old nevocellular nevi, sebaceous nevus, neurofibromas, connective tissue nevi, epidermal nevi, lipomas, acrochordons, focal dermal hypoplasia, lipoblastomatosis and Michelin tyre baby syndrome.
Histology reveals groups of ectopic mature adipocytes between the collagen bundles in the dermis with no connection of these adipocytes with the subcutaneous fat. Similar dermal collections of the adipose tissue may occur as a component of intradermal melanocytic naevi and in pedunculated lipofibroma.

To date, there have been no reports of malignant degeneration and recurrences, so treatment is not medically necessary. For cosmetic purposes, surgical excision is the best choice.
At birth this 11-year-old girl was noted to have a tumor on the left hip comprised of multi-lobular papules and nodules forming cerebriform plaques. The soft fleshy lesions were not symptomatic or associated with underlying defects of the underlying bony or soft tissue structures. 

(By Dr. Adem Koslu MD) - Dermatlas
This healthy adolescent complained of an irritating skin tag on her posterior thigh which had been slowly growing for 8-10 years. Histology demonstrated the typical changes of nevus lipomatosus. Contributor - Dr. David Cohen MD - Dermatlas
References

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare hamartomatous skin lesion histologically characterised by the presence of mature fat tissue within the dermis. Clinically, two types of NLCS can be distinguished: a multiple type of Hoffmann-Zurhelle and a solitary type. We report a retrospective study of 13 cases of NLCS seen in the Anatomopathological department of La Rabta hospital of Tunis during a period of 12 years (1992-2004). Two clinical forms were distinguished: the solitary form (11 cases) consisting of a unique papulo-nodular lesion and the multiple form (2 cases). Histologically, the tumor consisted in all cases on mature fat tissue.

PECULIARITIES IN DERMATOLOGY. A CASE OF NAEVUS LIPOMATOSUS SUPERFICIALIS (ZURHELLE) - KINGSLEY HJ.
Rare by definition is described as **uncommon, infrequent or of uncommonly high quality**. In our professional careers, irrespective whether in Private Practice or in Teaching Institutions, we will encounter clinical conditions which belong to such a definition and it will be up to us to recognize them and whenever possible to report them as well!
THANK YOU

Rare Disease Day
Blaschko's lines are an extremely rare and unexplained phenomenon of human anatomy first presented in 1901 by German dermatologist Alfred Blaschko. Neither a specific disease nor a predictable symptom of a disease, Blaschko's lines are an invisible pattern built into human DNA. Many inherited and acquired diseases of the skin or mucosa manifest themselves according to these patterns, creating the visual appearance of stripes. The cause of the stripes is thought to result from mosaicism; they do not correspond to nervous, muscular, or lymphatic systems. What makes them more remarkable is that they correspond quite closely from patient to patient, usually forming a "V" shape over the spine and "S" shapes over the chest, stomach, and sides.