A Bluish Pigmented Cystic Lesion of the Nose: A Quiz

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A 63-year-old Caucasian woman presented with a pigmented lesion of the left ala of the nose, which had been present for 6 years. Her medical history included diabetes, hypertension, hypercholesterolaemia, Laugier-Hunziker disease and rosacea. The lesion had not evolved in size, but had tended to darken progressively. There was no familial history of any specific cutaneous disorder. She had not noticed any triggering factors. Examination showed a translucent intense dark-blue pigmented lesion, 2 × 2 mm in diameter (Fig. 1a). The patient had no similar lesions elsewhere. 3 months later the lesion had not changed in size or colour. Dermatoscopy revealed a bluish nodule surrounded by several telangiectasias. Because the patient showed fear of skin cancer, a 4-mm punch skin biopsy was performed. The biopsy specimen disclosed a normal epidermis. The deep dermis contained a solitary cyst lined with a single or double layer of cuboidal epithelial cells with an eosinophilic cytoplasm. Extravasation of red blood cells was noted within the cystic cavity. No apocrine decapitation secretion or myoepithelial lining outside the cyst was observed (Fig. 1b).

What is your diagnosis? See next page for answer.

Asymptomatic Brownish Reticulate Patch on the Left Thigh: A Quiz

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A healthy 29-year-old woman presented with an asymptomatic, mildly erythematos to brownish reticulate patch, which had been present for a few months, on the anterior face of her left thigh (Fig. 1). The lesion was fixed, and was not blanchable, tender or migrating.

The patient denied any contact with chemicals, acute thermal exposure or repeated frictional trauma. She reported that the lesion had developed within a few months after she had started using a laptop computer for working on her thesis. No laboratory abnormalities were found. Histological investigations were not performed.

What is your diagnosis? See page 557 for answer.

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Fig. 1. (a) Clinical presentation. (b) Unilocular cyst of the mid-dermis with no communication with the epidermis (haematoxylin and eosin (H&E) 2). (c) Close-up view: single cystic cavity composed of 1 or 2 layers of cuboidal cells in the mid-dermis. (H&E 10). (Charlotte Pernet provided the pictures)
A 5-year-old Caucasian girl presented with a 6-month history of asymptomatic macules on her cheeks (Fig. 1). She had been treated with terbinafine hydrochloride 1% cream, hydrocortisone butyrate 0.1% cream and a course of amoxicillin-clavulanic acid with no improvement. Clinical examination showed multiple, scattered, rounded 2- to 4-mm in diameter macules localized on the cheeks. The lesions had regular orange-brown pigmentation. No other mucosal or cutaneous lesions were noted, and the child was otherwise well with no relevant past medical history. In particular, she had no constitutional symptoms and no evidence of lymphadenopathy or hepatosplenomegaly.

A complete blood count, erythrocyte sedimentation rate, a comprehensive metabolic panel, abdominal ultrasound scan and systemic skeletal radiographic survey showed no abnormalities. A skin biopsy was performed for histological examination with haematoxylin and eosin staining (Fig. 2A), integrated with immunohistochemical staining with CD68 (Fig. 2B), S-100 (Fig. 2C) and CD207 (Lang-erin, Fig. 2D).

**What is your diagnosis?** See page 558 for answer.

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**A Bluish Pigmented Cystic Lesion of the Nose: Comment**


**Diagnosis:** Solitary (“Smith and Chernovsky” type) eccrine hidrocystoma

Eccrine hidrocystoma (EH) is a benign cystic tumour of the eccrine duct. It may present in two distinct ways. The “classic” Robinson type, described in the end of the 19th century (1), is characterized by multiple cystic lesions of the face of middle-aged women who work in hot environments. Lesions tend to enlarge and become more symptomatic in the summer or in case of exposure to hot/moist air. However, this type seems to be less frequent nowadays due to the development of cooler working conditions (2, 3). The “Smith and Chernovsky” type, described 80 years later, refers to solitary (usually less than four) facial lesions, affecting both women and men. Lesions do not display any seasonal variation. EH are mostly located on the periorbital areas, but lesions may be found on other areas of the face, head, trunk and popliteal fossa (2). Localization on the nose is rather unusual (3, 4). Individual EH present as asymptomatic, smooth, shiny, flesh-coloured to various shades of blue, dome-shaped papules ranging from 1 to 16 mm in diameter. Rupture of the cyst, resulting in collapse of the lesion with the loss of a watery fluid, helps to confirm the diagnosis (2). Differential diagnoses include apocrine hidrocystoma (AH) and basal cell carcinoma (2). Distinction from AH is difficult: AH is reported to be larger, darker blue and less likely to be periorbital compared with EH, as in our case (2). The final diagnosis is made by the pathologist: lining epithelium consisting of secretory and myoepithelial cells, S-100 protein negative staining, decapitation of secretory cells, and Periodic acid-Schiff (PAS)-positive granules support a diagnosis of EH (3). Cancerophobia, as in our case, or cosmetic consequences may result in a biopsy or excision of the lesion being carried out (2). Dermatoscopy may be useful in distinguishing EH from basal cell carcinoma (5).

**Diagnosis:** Erythema ab igne caused by use of a laptop computer on the lap.

Erythema ab igne (EAI) is a well-known adverse effect of repeated long-term exposure to mild heat in the range 43–47°C, which is insufficient to cause a burn (1, 2).

Clinically, EAI is typically characterized by the presence on the skin of an asymptomatic, fixed, non-tender, brownish-red, reticulate patch, which is variable in shape and width.

In the early stages, histopathological changes observed in EAI include epidermal atrophy, dermal pigmentation and vasodilatation. Focal hyperkeratosis and epithelial cellular atypia occur later, closely resembling the changes induced by actinic damage (3). Melanocytes with increased dendritic processes and abundant melanophages are present in the dermis. There may also be an accumulation of dermal elastic tissue, which is an early sign of both ultraviolet (UV) radiation and heat-induced skin damage (4). The similarities between EAI and actinic keratoses suggest that heat may induce epithelial changes as a result of clonal mutation in the same way that UV light produces epithelial changes (5).

Squamous cell carcinoma and neuroendocrine carcinoma, also known as Merkel cell carcinoma, may arise in the site of EAI on rare occasions (1, 5). The most common thermally induced cancer, squamous cell carcinoma, tends to occur after a period of more than 30 years (4, 6).

The most important differential diagnosis is between EAI and livedo reticularis, but there are significant clinical and anamnetical differences. A biopsy should be performed if there is any evidence of cutaneous malignancy or in cases of diagnostic doubt.

In the past, EAI was usually seen on localized areas of the body that were closely and repeatedly exposed to heat, normally from fireplaces, braziers, wood-burning stoves, steam radiators and space heaters (2, 5). Since the widespread availability of central heating the incidence of EAI has decreased (6), but new and often unusual causes have been described in the literature. These include hot water bottles and heating pads used to treat stomach-ache, muscular and arthritic pain, or various other chronic pains (4), furniture with a built-in heating unit (7), heating blankets (8), car heaters (9), hot bathing (2), heated popcorn (10) and a sauna belt system used to treat abdominal obesity and cellulite (11).

Several cases of EAI induced by laptops have been reported to date since the first publication in 2004 (12). This number is likely to increase with time, as laptops are used increasingly (5, 6, 12–14). In this case, the heat source causing EAI is the battery in the base of the laptop and the classical skin area involved is the thighs. EAI therefore may become an occupational hazard not only for bakers, foundry workers and kitchen workers, but also for people working with their laptops on their thighs (4, 14). A further problem for laptop users is the risk of a negative influence on fertility (15).

The mainstay of treatment of EAI is immediately to remove the source of infrared radiation; in which case the prognosis of EAI is excellent (4, 6, 14). Residual hypo-pigmentation or hyper-pigmentation may be observed, especially in dark skin types.

In the present case the patient reported that she worked every day with the laptop placed directly on her thighs for at least 3–4 h. The unilaterality of the lesion on the left thigh is explained by the location of the battery on the bottom left-hand side of the laptop, which is subjected to warming. The patient was instructed to avoid direct contact with the laptop or to use an insulating material between her skin and the computer. After 3 months the patch of EAI was significantly improved.

REFERENCES


Multiple Brownish Macules on Child’s cheeks: Comment
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Diagnosis: Indeterminate cell histiocytosis

Biopsy specimen from a lesion showed a dense upper dermal infiltrate of large, round-to-ovoid, histiocytes characterized by kidney-shaped or oval nuclei and an ill-defined cytoplasm, with few admixed lymphocytes. No significant eosinophilic or plasma cells infiltration were noted. Neoplastic cells were situated immediately beneath a thinned epidermis devoid of rete ridges and were strongly positive for CD-68, positive for S-100 and negative for CD-207. Moreover, no epidermotropism, xanthomatized histiocytes or Touton giant cells were observed.

The girl’s parents declined a further skin biopsy for an electron microscopy evaluation and no therapy was prescribed. After 4 years the macules have diffused all over the body and some have developed into fleshy, dome-shaped papules. No systemic involvement has been noted, as documented by extensive clinical, laboratory and radiological follow-up.

DISCUSSION

Indeterminate cell histiocytosis (ICH) is a rare disorder characterized by the proliferation of histiocytic cells that express S-100 antigens, but, unlike Langerhans’ cell histiocytosis (LCH), lack Birbeck granules. The patient reported here presented with yellow-orange macules confined to the face without visceral involvement. It should be noted that the clinical picture, with the exception of the age of patients and of the lack of regression of cutaneous lesions, shares striking similarities to that of benign cephalic histiocytosis, which represents the sole clinical differential diagnosis. Histopathological differential diagnosis includes LCH, juvenile xanthogranuloma, benign cephalic histiocytosis and the cutaneous variant of Rosai-Dorfman disease (CRDD). Besides clinical considerations, LCH can be excluded because of the lack of epidermotropism, the scarcity of eosinophils infiltrate and the absence of Birbeck granules, as documented by negativity of the CD-207 (1). Histiocytes in juvenile xanthogranuloma are vacuolated, Touton cells can be observed in the infiltrate and, finally, histiocytes are, by definition, S-100 negative; the latter feature also characterizes BCH, which can therefore be readily excluded; unlike in our case, the infiltrate in CRDD has typically a nodular appearance with emperipolisis and karyophagocytosis and presents a dense collection of plasma cells.

ICH usually presents with solitary (2, 3) or multiple asymptomatic, flesh-coloured to yellow-brown maculopapules with no site of predilection. According to the literature the clinical appearance of this case resembles one of the variants of NLCH (4). Although ICH has been described in childhood (2, 5) the mean age reported in a recent large case series was 46 years (4). In general, ICH has a good prognosis, being mostly exclusively a cutaneous disorder, but a fatal paediatric case has been described (6). Various anecdotal treatment has been described as effective for this condition, including pure coal tar and 5% 5-fluorouracil cream (5), narrow-band ultraviolet B (7) and psoralen plus ultraviolet A phototherapy (8). Moreover, acute myeloblastic leukaemia was reported following chemotherapy for ICH (9). Taking into consideration the young age of our patient, and the presence of few cutaneous lesions limited to the face with no visceral involvement, we have chosen to not treat the child, but to monitor her clinical course closely, deferring therapy for potential progression of the disease.

REFERENCES