This month – 6 cases:

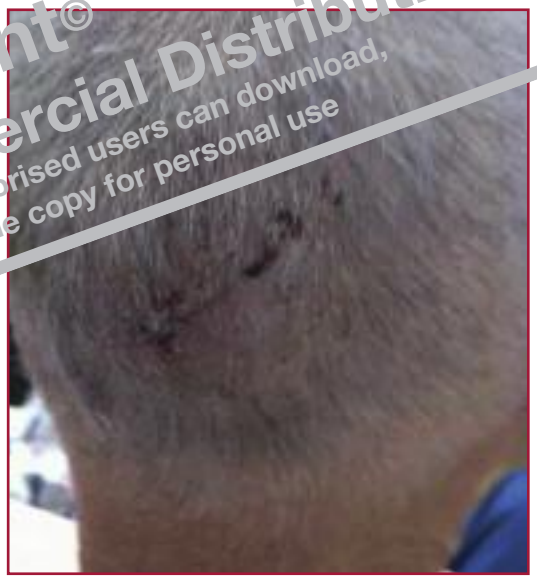
Case 1

Soft Tissue Mass on the Occiput

An 80-year-old male has developed a progressively growing soft tissue mass on his occiput over a six month period of time. It has become painful, making lying supine difficult.

What is your diagnosis?

a. Sebaceous cyst
b. Lipoma
c. HCC metastasis to the skull
d. Exostosis



Answer

Hepatocellular carcinoma (HCC) metastasis to the skull **(answer c)** – biopsy proven – is the diagnosis. HCC is an aggressive tumour. It is the fifth most frequently diagnosed cancer, but the second leading cause of cancer-related death in men. The majority of HCCs occur in patients with chronic liver disease.

The most common sites of HCC metastasis are the lung, intra-abdominal lymph nodes, bone, and the adrenal gland. Brain involvement is rare.

It is always prudent to consider metastasis as a differential diagnosis when a patient with malignancy presents with a mass or lesion somewhere else.

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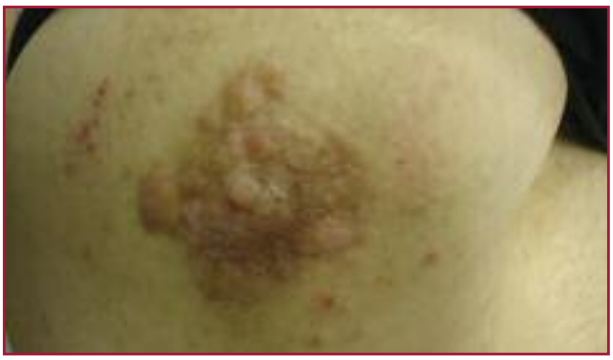
Kathy O'Connor, RN (EC), is a Nurse Practitioner with the North Renfrew Family Health Team, Deep River, Ontario.

Case 2

Enlarged Lesion on the Thigh

This 60-year-old man first noted a lesion on his thigh 20 years ago. It has enlarged over time with colour and textural changes. It remains asymptomatic.



What is your diagnosis?

a. Keloid
b. Dermatofibrosarcoma protuberans
c. Hemangioma
d. Melanoma
e. Dermatofibroma

Answer

Dermatofibrosarcoma protuberans (DFSP) **(answer b)** usually begins as an asymptomatic skin-toned papule that gradually progresses to a more indurated plaque or nodule with variable colour changes.

It most commonly affects young adults. It favours the trunk in 60% of persons, proximal extremities in 30%, and the head and neck in 10%.

While DFSP is locally invasive, it may extend deeply into subcutaneous tissue; hence, it is prone to recurrence.

A wide Mohs surgical excision is favoured.

Stanley Wine, MD, FRCPC, is a Dermatologist in North York, Ontario.

Case 3

A Hairless Patch

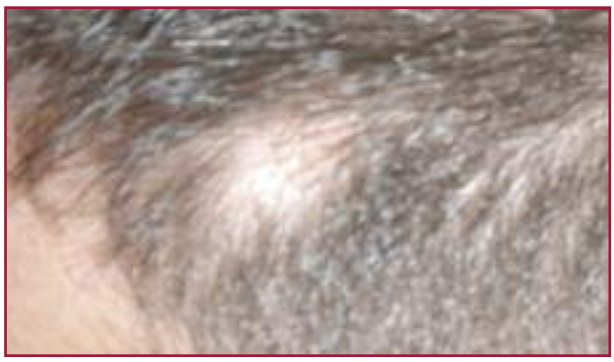
This man consulted his doctor about a hairless patch that he has had for as long as he can remember. His younger brother has a similar patch. He is concerned that he may be going bald.

What is your diagnosis?

a. Temporal triangular alopecia
b. Alopecia areata
c. Trichotillomania
d. Moth-eaten alopecia

Answer

Temporal triangular alopecia (TTA) **(answer a)** presents as oval or triangular areas of alopecia in the temporal areas of the scalp. Onset is usually between two– to six-years-of-age. Bilateral lesions may occur. The "bald spot" actually contains a normal amount of hair, but the hairs are vellus, rather than terminal.



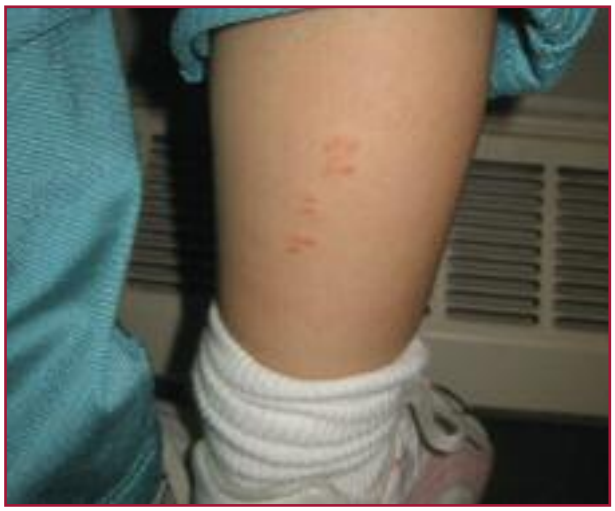
No treatment is usually needed; however, complete surgical excision or hair transplantation may be performed.

Hayder Kubba, MBChB, LMCC, CCFP, FRCS (UK), DFFP, DPD, graduated from the University of Baghdad, where he initially trained as a Trauma Surgeon. He moved to Britain, where he received his FRCS and worked as an ER Physician before specializing in Family Medicine. He is currently a Family Practitioner in Mississauga, Ontario.

Case 4

Linear Erythematous Leg Papules

A two-year-old girl presents with a two month history of linear erythematous papules over the lower leg.



What is your diagnosis?

a. Flat warts
b. Incontinentia pigmentia
c. Linear morphea
d. Lichen striatus
e. Herpes zoster

Answer

Lichen striatus **(answer d)** is a unilateral dermatitis that is self-limiting, peaking in severity in weeks to months. It has a female predilection. It presents as small pink, tan, or skin-coloured papules in a linear band distribution following the developmental lines of Blaschko. A fine silvery scale may overlie these papules. In darker-skinned individuals, this scale is more prominent; there may also be an area of hypopigmentation. Puritus may be present.

No treatment is necessary, as these lesions will spontaneously resolve in 3 to 12 months. There may be residual hypopigmentation following resolution.

Incontinentia pigmenti (Bloch-Sulzberger syndrome) is an X-linked dominant condition that is lethal in males. In addition to the skin, it affects the eyes, CNS, and teeth. Skin lesions tend to follow the lines of Blaschko and progress through four phases: vesicular, verrucous, hyperpigmented, and hypopigmented/atrophic. Our patient did not have these skin lesions at birth, making this diagnosis less likely.

Linear morphea is a type of scleroderma that is localized to skin. Patients may have arthralgias, but lack systemic involvement, such as sclerodactyly or Raynaud's syndrome. These lesions usually do not follow the developmental lines of Blaschko. Our patient's lesion is not band-like and does not follow the entire length of the extremity, thus making linear morphea less likely.

Flat warts, or verruca plana, are benign intraepidermal tumours of skin mucosa overgrowth caused by the human pappilomavirus. They are small, smooth, skin-coloured papules with flat tops. There may be a single to hundreds of warts that can join together to form a plaque. Our patient's papules are more erythematous than the typical wart.

Herpes zoster, or shingles, is caused by a reactivation of a latent varicella zoster virus (VZV) infection. The lesions are typically distributed along a dermatome unilaterally. The lesions are painful vesicles on an erythematous base. It is more common in the elderly or immunocompromised. Our patient does not have vesicular lesions, which makes this diagnosis less likely.

Joseph M. Lam, MD, is a Pediatrician with fellowship training in Pediatric Dermatology. He practices in Vancouver, British Columbia.

Karen Chu is a final year medical student at the University of British Columbia, Vancouver, British Columbia.

Case 5

Hyperpigmented Leg Lesions

A 25-year-old man presents with hyperpigmented lesions on his legs. The lesions were first noted during early childhood. His 48-year-old father and 20-year-old brother have similar lesions on their legs.

What is your diagnosis?

a. Lamellar ichthyosis
b. Nonbullous congenital ichthyosiform erythroderma
c. Ichthyosis vulgaris
d. Bullous congenital ichthyosiform erythroderma

Answer

Ichthyosis vulgaris **(answer c)** is an autosomal dominant disorder of cornification. The primary genetic defect is a nonsense mutation in the filaggrin gene (*FLG*), resulting in reduced filaggrin expression. The lesions are not usually present at birth but appear in most patients during the first year of life and in the vast majority by age five. The scaling is symmetric and usually intensifies until puberty, and subsequently decreases with age. The colour of the fine, fish-like scales varies from white to dirty gray to brown. Scaling is most prominent on the extensor aspects of the extremities, particularly the shins.

Lamellar ichthyosis, an autosomal recessive disorder, usually presents at birth with a parchment-like collodion membrane, (hence the term "collodion baby") that desquamates over the next 10 to 14 days. Over time, large, dark brown, plate-like scales develop. These are centrally adherent with raised edges and resemble a suit of armour. Tautness of the facial skin might result in eversion of the eyelids (ectropion) and lips (eclabium).

Nonbullous congenital ichthyosiform erythroderma, an autosomal recessive disorder, also presents at birth with a collodion membrane. After shedding of the membrane, pronounced erythroderma and fine white scales distinguish this condition from other forms of ichthyosis.

Bullous congenital ichthyosiform erythroderma, an autosomal dominant disorder, usually presents at birth with erosions, large areas of denuded skin, and erythroderma. Over time, blistering and erythroderma diminish and hyperkeratosis develops. Thick brown scales cover most of the skin surface, especially in the flexural areas.

Alexander K.C. Leung, MBBS, FRCPC, FRCP(UK&Irel), FRCPCH, is a Clinical Professor of Pediatrics, at the University of Calgary, Calgary, Alberta.

Andrew Wong is a Family Medicine Resident at the University of Calgary, Calgary, Alberta.

Case 6

Itchy, Evanescent Lesions

This patient presented with these itchy lesions that come and go within a few hours. He has noticed that they are triggered by spicy foods he enjoys.

What is your diagnosis?

a. Urticaria
b. Pemphigoid
c. Pityriasis rosea
d. Acne vulgaris

Answer

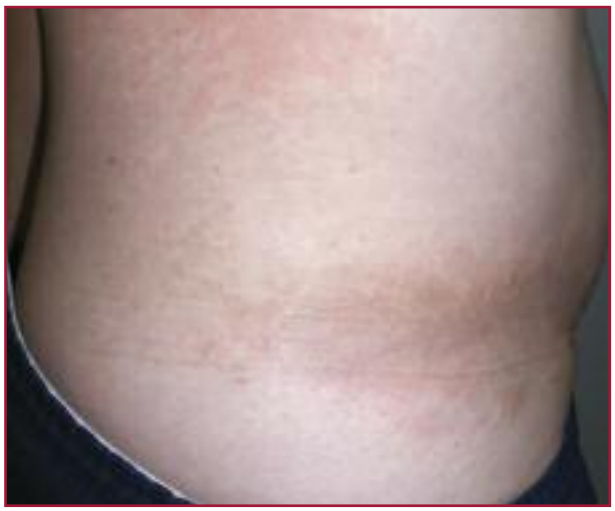
Urticaria **(answer a)** is pruritic. The degree of pruritus depends on the amount of swelling. Patients often say they see the lesions come and go within hours. The lesions are evanescent, migratory, and do not usually last beyond 24 hours. Urticaria presents as elevated and edematous plaques, usually pale, red, or pink. As the lesions spread, there is often central clearing, which leaves an arcuate or gyrate formation.

Urticaria may be acute or chronic. By definition, acute urticaria lasts less than six weeks, and chronic urticaria lasts longer. The causes of either are rarely found. Common precipitants include foods, drugs, vitamins, viral infections, bacterial infections, helminth infections, bites or stings, and vitamins.

Food allergens have long been considered to be potential causes of urticaria. The exact incidence varies from 2 to 30% but is more likely to be in the lower range. Common potential offenders include fruits, nuts, eggs, soya beans, wheat, fish, seafood, tea, and food additives, such as aspartame.

Urticaria can also be caused by physical agents, such as heat, cold, ultraviolet light, and trauma from scratching (dermatographism). Exercise-induced urticaria (cholinergic urticaria) produces small lesions 1 to 2 mm in diameter.

Diagnosis of urticaria can often be made purely based on symptoms and signs. Dermatographism can be elicited by lightly scratching the skin. When the diagnosis is not certain, a punch biopsy may help.



The premise is that histamines are the major physiologic agents inducing urticaria. Since the vasculature has both H1 and H2 receptors, it is important to consider the use of both types of antihistamines. The caveat with most antihistamines is adequate dosing. Keep in mind that the recommended dose is usually for a 70 kg patient. In our affluent, well-fed society many patients are at least 50% over the average weight.

For mild, limited disease, withdrawing the causative agent may be all that is needed. In addition, oral H1 blockers may be helpful. Finally, a short course of an oral corticosteroid tapered over 10 days may be needed. A good beginning dose is prednisone 1 mg/kg q.d.

Patients should always understand that urticaria, acute or chronic, is a serious symptom complex that may not respond to therapy and that oral corticosteroid treatment to prevent serious airway complications may be necessary for prolonged periods of time. However, most urticaria clears with therapy without discovering a cause.

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