This month – 6 cases:

Case 1

Waxing and Waning Lesions

A six-year-old girl broke out with an itchy rash all over her body two hours ago. The lesions wax and wane rapidly.

What is your diagnosis?

a. Urticaria
b. Mastocytosis
c. Erythema multiforme
d. Bullous pemphigoid

Answer

Urticaria **(answer a)** is characterized by pruritic, erythematous, edematous wheals of the superficial layers of the skin. The hallmark of urticaria is that individual lesions wax and wane rapidly, usually lasting less than four hours and rarely persist longer than 24 hours. The lesions blanch on pressure, vary in size from a few millimeters to a few centimeters in diameter and can be localized or generalized. The lesions are usually well circumscribed with central clearing. Peripheral extension and coalescence of individual lesions result in a clinical picture of oval, annular, or bizarre serpiginous configurations.

Triggering factors should be avoided if possible. Currently, the most frequently used therapy aims at blocking the release of mediators from mast cells or blocking the effects of released mediators.

Non-sedating H1 antihistamines are the mainstays in the management of urticaria. In acute severe urticaria, subcutaneous epinephrine (0.01 ml of 1:1,000 epinephrine per kg, up to 0.3 ml) is indicated.

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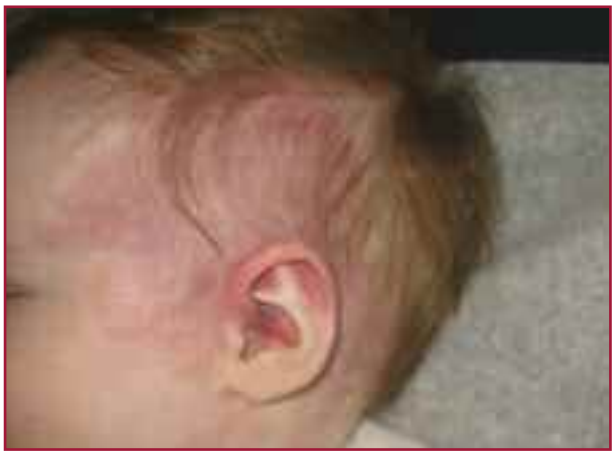
Case 2

A Unilateral, Pink Lesion

A six-month-old infant presents with a unilateral, pink patch along the left side of her head sparing most of her forehead and orbit areas. It has been present since birth and remained static without any thickening. She appears otherwise healthy with no symptoms, however her parents remain particularly concerned about the possibility of neurological symptoms arising.



What is your diagnosis?

a. PHACES syndrome
b. Port-wine stain
c. Mongolian spot (dermal melanocytosis)
d. Klippel-Trenaunay syndrome
e. Nevus simplex (angel's kiss)

Answer

Port-wine stain (PWS) **(answer b)**, or nevus flammeus, is a congenital capillary malformation which can occur as an isolated lesion or in association with a number of syndromes. The lesions are macular stains that range from pink to dark red in colour. Over time, these lesions may darken and become hypertrophied. PWS is usually unilateral and most often affects facial sites although they can occur on any cutaneous surface. The major significance of the PWS revolves around their potential cosmetic impact on the developing child and their potential for syndromic associations. In this case, the possibility of Sturge-Weber syndrome should be considered.

PHACES association is an acronym to describe a rare neurocutaneous syndrome that can include posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, eye abnormalities and sternal defects. PHACES syndome is associated with hemangiomas of infancy and not with PWS.

Mongolian spots represent collections of spindle-shaped melanocytes located deep in the dermis and present as deep brown to slate gray or blue-black macular lesions that are usually poorly circumscribed and typically seen in non-Caucasian populations.

Klippel-Trenaunay syndrome is a sporadic disorder distinguished by the triad of vascular malformation, venous varicosity and hyperplasia of soft tissue and bone. The most common location to be involved is the lower extremity with the various lesions also usually distributed on that same extremity.

A nevus simplex is a dull pink macular lesion that represents the most common vascular lesion of infancy. These lesions are accentuated during periods of crying, breath-holding, or physical exertion. Unlike PWS, the lesions on the face commonly fade within the first two years of life.

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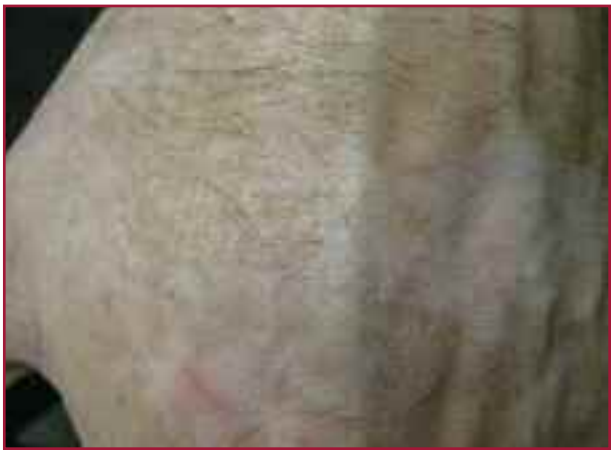
Case 3

Worrisome White Patches

A 38-year-old Caucasian letter carrier presents with white patches on his hands and around his mouth. They are asymptomatic and he is worried he may have a fungal infection.



What is your diagnosis?

a. Tinea versicolor
b. Pityriasis alba
c. Chemical leukoderma
d. Vitiligo
e. Piebaldism

Answer

Vitiligo **(answer d)** is an acquired, progressive leukoderma with depigmentation of the epidermis. Melanocytes are destroyed in the skin and those in hair follicles can also be affected resulting in white hair. The cause of this condition is not fully elucidated, but there is a role for an inherited disposition for vitiligo. Approximately 1% to 2% of the population is affected and there is familial clustering of cases. While vitiligo can develop at any age, typically the onset is at 10- to 30-years-of-age.

Vitiligo progresses without symptoms, though early lesions may be pruritic. Vitiligo presents as sharply demarcated patches that are cosmetically disturbing to most patients, especially those with darker skin colour. After a few small white lesions, they increase in number and size, becoming confluent and having unusual shapes. Vitiligo is classified as localized (*e.g.*, segmental) or generalized involvement. The most commonly affected areas are the face, neck and scalp. Areas of repeated trauma are also prone, including bony prominences, dorsal hands and ventral wrists.

Treatment of this condition is unsatisfactory in many cases. Potent topical steroids and topical calcineurin inhibitors (*e.g.*, tacrolimus) are often employed initially and phototherapy employed for widespread or resistant cases.

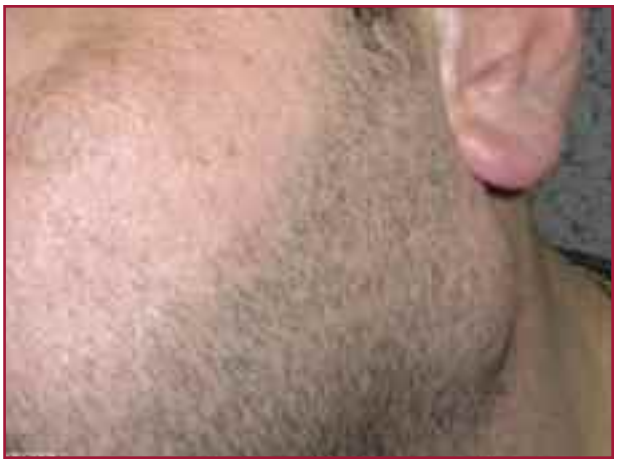
Benjamin Barankin, MD, FRCPC, is a Dermatologist practicing in Toronto, Ontario.

Case 4

Neck Swelling

This 45-year-old gentleman presents with this painless neck swelling, which he has had for the last three years or so. He did not come earlier since he was very concerned about the possibility of cancer as his mother had a permanent colostomy about two years ago for a very low rectal cancer.

He works as a truck driver, does not smoke and drinks 2-3 units of alcohol a day. He is otherwise well, did not notice any recent weight changes and denies any fever, itching or night sweats. His examination revealed a slightly overweight man with no sign of pallor. He had neither lymphadenopathy nor hepatosplenomegaly.



What is your diagnosis?

a. Benign lymphoepithelial lesions of parotid gland
b. Lymphoma
c. Mumps
d. Warthin's tumour
e. Pleomorphic adenoma

Answer

Pleomorphic adenoma **(answer e)** of the salivary glands is also known as a benign mixed tumour and presents as a painless mass, typically in adults during their third to fifth decades. The tumour has circumscribed appearance when it is cut.

The majority of these tumours arise within the parotid gland (70% to 85%). When occurring in minor salivary glands, the palate is the most common site (60% to 65%).

Complete surgical removal is curative but if the initial surgical procedure does not completely remove the tumour, there is a low recurrence rate < 2%.

Rarely, a malignant tumour may arise within this tumour, a phenomenon known as carcinoma ex pleomorphic adenoma. This has been reported to occur in 2% to 7% of cases.

The most reliable features to determine malignancy include an infiltrative growth pattern, vascular permeation, perineural invasion and marked cytological atypia with abnormal mitotic figures.

Mumps nowadays is not a common cause for parotid enlargement because of immunization and usually is associated with systemic illness.

Warthin's tumour is a benign neoplasm of the salivary gland. An older name is papillary cystadenoma lymphomatosum. It accounts for 4% to 5% of salivary gland neoplasm and is more common in men during their sixth to seventh decades. It almost exclusively occurs in the parotid gland and is bilateral in 5% of cases. The tumour presents as a painful swelling, usually within the lower portion of the salivary gland.

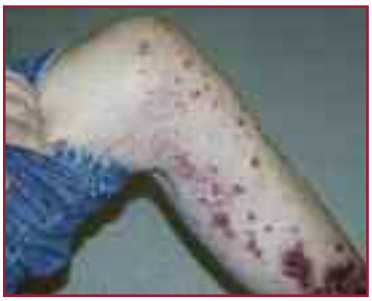
Benign lymphoepithelial lesions of parotid gland is usually bilateral.

Hayder Kubba 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 5

Erythematous Spots

An 83-year-old male developed malaise, myalgia and joints pain, followed by non-itchy erythematous spots on his lower and upper extremities, abdomen and chest. A few days before the rash, he started a herbal medication for his problems with prostate.



What is your diagnosis?

a. Erythema nodosum
b. Ecchymoses
c. Vasculitis
d. Meningococcal septicaemia

Answer

Vasculitis **(answer c)** is the inflammation of blood vessels. It has a host of manifestations depending on the size, type and location of the vessels affected. It is not a diagnosis in itself, but part of a range of pathological processes.

Many tissue can be affected, but the skin, because of its profuse vascular supply and ready visibility, is often first recognized, usually on the lower legs, but otherwise patients might therefore present with a wide variety of clinical signs and symptoms. Unfortunately, no clinical pattern is specific to any underlying cause.

Identify and remove the offending antigen. Short courses of prednisone (40 mg to 60 mg q.d.) may be very useful for patients with severe symptoms.

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Case 6

A Scaly Plaque

A 43-year-old male from Ethiopia who recently immigrated to Canada, presents with a six month history of annular scaling plaques on the trunk, arms and legs. The lesions did not respond to treatment with topical antifungal creams.

What is your diagnosis?

a. Cutaneous lupus erythematosus
b. Psoriasis
c. Nummular dermatitis
d. Tinea corporis

Answer

This patient had fungal cultures positive for *T. soudanense* confirming a diagnosis of Tinea corporis **(answer d)**. Tinea corporis, also known as "ringworm," is a dermatophyte infection of the skin. Tinea infections are named according to the area of the body infected. For example, Tinea manuum, Tinea faciei and Tinea capitis affect the hands, face and scalp, respectively. Symptoms can include itching and burning at the site of a scaly, annular lesion with a raised active border. Dermatophytes are not endogenous pathogens and the fungus has an incubation period of one to three weeks. The infection spreads from human to human, animal to human and soil to human contact.

T. rubrum and *T. mentagrophytes* are common pathogens that cause Tinea corporis. *T. soudanense* is more commonly found in parts of Africa, however, increased international immigration and tourism will likely result in more dermatophyte infections of this kind in Canada.

Localized infections may be treated with topical antifungals (*i.e.*, clotrimazole), however, more extensive infections may require treatment with a systemic antifungal (*i.e.*, terbinafine, itraconazole or fluconazole).

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