**This month – 8 cases:**

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**Case 1**

## Yellowish-Orange Back Papule

A nine-month-old female presents with an asymptomatic yellowish-orange papule on her back. She is otherwise healthy.

### What is your diagnosis?

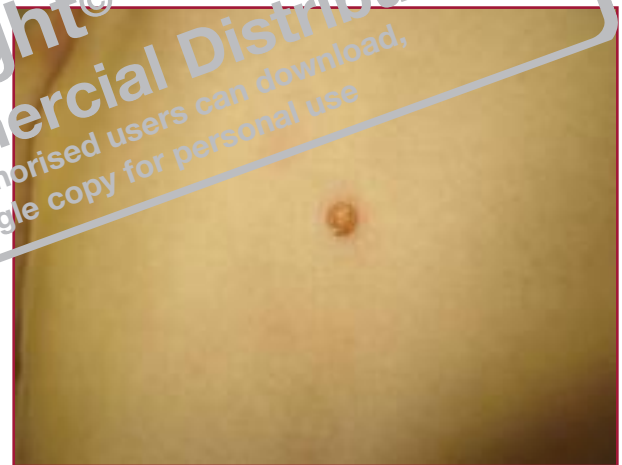
- a. Juvenile xanthogranuloma
- b. Spitz nevus
- c. Dermatofibroma
- d. Dysplastic nevus
- e. Solitary mastocytoma

### Answer

A juvenile xanthogranuloma (JXG) (**answer a**) is a benign, asymptomatic, usually self-resolving, orange-to-yellow papule composed of histiocytic cells that predominantly occur in infancy and childhood.

Papules occur most commonly on the skin, but rarely can affect the eyes and viscera. A skin biopsy can be performed if the diagnosis is in doubt, and/or unless multiple JXGs are present. Further workup is

seldom required. Skin JXGs can be excised for cosmetic reasons, or in most cases simply observed, as many do resolve on their own.



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

**Case 2**

# *Muscle Atrophy and Dry Cough*

A 57-year-old male presents with a recurrent dry cough. During examination muscle atrophy of the right arm and forearm with weakness were found. He stated that he developed such a problem during childhood. His condition is stable now. His medical history, physical examination and clinical investigations are otherwise unremarkable.

## *What is your diagnosis?*

- a. Right brachial plexopathy
- b. Herniated cervical disc
- c. Poliomyelitis
- d. Cervical spinal stenosis

## *Answer*

Poliomyelitis (answer c), often called polio or infantile paralysis, is an acute viral infectious disease spread from person to person, primarily via the fecal-oral route. Polio follows infection with any one of three related enteroviruses – poliovirus types 1, 2, or 3. The virus enters through the mouth and then multiplies inside the throat and intestines. The incubation period is 4 to 35 days and the initial symptoms include fever, fatigue, headaches, vomiting, constipation (or less commonly diarrhea), stiffness in the neck, and pain in the limbs. Although around 90% of polio infections cause no symptoms at all, affected individuals can exhibit a range of symptoms if the virus enters the blood stream. In about 1% of cases, the virus enters the central nervous system, preferentially infecting and destroying motor neurons, leading to muscle weakness and acute flaccid paralysis.

Different types of paralysis may occur, depending on the nerves involved. Spinal polio is the most common form, characterized by asymmetric paralysis that most often involves the legs. Bulbar polio leads to weakness of muscles innervated by cranial nerves. Bulbo-spinal polio is a combination of bulbar and spinal paralysis. Around a quarter of individuals who survive paralytic polio in childhood



develop additional symptoms decades after recovering from the acute infection, notably muscle weakness, extreme fatigue, or paralysis. This condition is known as post-polio syndrome (PPS). Not everyone who has had polio will develop late effects. People who have been severely paralyzed by polio are most commonly affected. Approximately 20 to 40% of people who had acute paralysis due to polio will develop these late effects.

The symptoms of PPS are thought to involve a failure of the oversized motor units created during recovery from paralytic disease. Factors that increase the risk of PPS include the length of time since acute poliovirus infection, the presence of permanent residual impairment after recovery from the acute illness and both overuse and disuse of neurons. Europe was declared polio-free in 2002, because of extensive vaccination programmes over many years. However, polio is still endemic in India, Pakistan, Nigeria and Afghanistan.

There is no cure for polio. Treatment is entirely symptomatic. Moist heat is coupled with physical therapy to stimulate the muscles and antispasmodic drugs are given to produce muscular relaxation.

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### Case 3

## *Tattoo Gone Awry*



This 35-year-old male had a tattoo placed on his shoulder one month ago. After three weeks he noted a papular rash along the tattoo lines

### *What is your diagnosis?*

- a. Reaction to black dye
- b. Pyoderma
- c. Atypical mycobacteria
- d. Koebner phenomenon

### *Answer*

The development of lesions in the line of prior trauma is an isomorphic response known as the Koebner Phenomenon (**answer d**). It is commonly seen with psoriasis, verruca plana, and in this case lichen planus. The individual lesions along the line of trauma are those typical of lichen planus (*i.e.*, small polygonal-shaped violaceous flat topped papules).

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

**Case 4**

## *Multiple Bluish Spots*

A three-month-old girl is noted to have multiple bluish spots on the back and buttocks. The spots have been present since birth.

### *What is your diagnosis?*

- a. Nevi of Ota
- b. Nevi of Ito
- c. Mongolian spots
- d. Multiple bruises

### *Answer*

Mongolian spots (**answer c**) are congenital hyperpigmented macules of varying size and shape. They are usually grayish in colour. Mongolian spots occur most commonly in the sacrococcygeal area, followed by the gluteal and lumbar areas. These lesions are less common on the back, and occur even less frequently on the abdomen, thorax, and limbs. Mongolian spots are usually round or ovoid. They are thought to be the result of dermal melanocytes failing to migrate to the epidermis. Mongolian spots are rare in Caucasians, but are very common in children of Asian and African descent. They usually fade during the first few years of life and are rare in children older than 10 years of age. Typical and limited Mongolian spots are benign skin markings and are not linked with any disorder. In rare instances an association is made between generalized Mongolian spots and inheritable storage disease, especially in children of Caucasian origin with generalized Mongolian spots.

A nevus of Ota is characterized as a benign melanosis of the skin around the eye, within the distribution of the ophthalmic and maxillary divisions of the trigeminal nerve. Pigmentation is usually unilateral and may involve ocular and oral mucosal surfaces.



In nevus of Ito, the pigmentation occurs in the acromioclavicular region and is more diffuse and less mottled. Unlike Mongolian spots, both nevus of Ota and nevus of Ito tend to persist through adult life.

A Mongolian spot can be distinguished from a bruise: a Mongolian spot does not change color and may take months to years to disappear. In addition, a Mongolian spot is usually homogeneous in color and is never tender. In contrast, a bruise is usually tender and tends to fade. A bruise has many more shades: the colour spectrum ranges from a blue-black or purple hue of the fresh injury to a yellow-green colour through the stages of hemosiderin breakdown.

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Carmen Fong is a medical student at the University of Calgary.

## Case 5

# Arm Papules



A 15-year-old female presents with concerns regarding the appearance of the back of her arms. The area is asymptomatic and her mother has similar lesions.

### *What is your diagnosis?*

- a. Squamous cell carcinoma
- b. Cutaneous horn
- c. Keratoacanthoma
- d. Infected molluscum contagiosum
- e. Nodular basal cell carcinoma

### *Answer*

Keratoacanthoma (KA) (**answer c**) is a low-grade malignancy that closely resembles a squamous cell carcinoma (SCC). Some pathologists now classify KA as a mild variant of invasive SCC. KA is characterized by rapid growth over a few weeks to months, followed by spontaneous resolution in many cases over several months.

Lesions typically present as solitary firm, round, skin-coloured, or reddish papules with a central crateriform keratin plug. Most KAs occur on sun-exposed areas.

Treatment is advocated because of the rare potential for invasion and metastases of lesions. KA can be treated by excision or electrodesiccation and curettage. Less commonly, radiation, cryotherapy or CO<sub>2</sub> laser can be used.

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

**Case 6**

## *Paediatric Papular Eruption*

A 14-month-old East Indian female presents with two skin coloured umbilicated papules over the back.

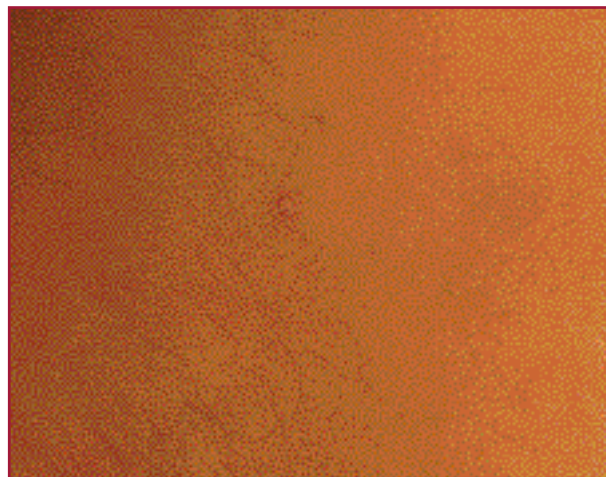
### *What is your diagnosis?*

- a. Pyogenic granuloma
- b. Langerhans cell histiocytosis
- c. Flat warts
- d. Lymphomatoid papulosis
- e. Molluscum contagiosum

### *Answer*

Molluscum contagiosum (MC) (**answer e**) is a common cutaneous viral infection in children caused by a member of the poxvirus. MC occurs most often in school-aged children, where it is easily spread through skin-to-skin contact and fomites. In affected patients, scratching or touching the lesions can spread the virus by autoinoculation. MC is characterized by firm, pearly, flesh or pink-coloured, dome-shaped papules. The papules range from 2 mm to 8 mm in size and often contain an umbilicated center. MCs are most common in moist areas, or areas where skin rubbing occurs, including the axillae, popliteal fossae, and groin. Genital and perianal lesions are also common. MC most often presents with numerous clustered papules, and linear configurations from koebnerization may be present. Surrounding dermatitis around MC lesions is common.

Complications of MC are rare, but include scratching-induced secondary bacterial infection. Spontaneous clearing of MC often occurs over years, but parents and patients may request therapy



for cosmetic reasons, pruritus, and epidemiologic concerns. Traditional therapies utilized in adults, such as curettage or cryotherapy, may be traumatic for pediatric patients. Many other methods of therapy have been reported for MC. One highly effective therapy is the in-office application of cantharidin, which is usually well tolerated when safety guidelines are followed.

Treatment results in blister formation within 24 to 48 hours, with healing over several days to one week. This agent is not generally recommended for treatment of perioral or periocular facial lesions, mucosal sites, or occluded areas such as the diaper region.

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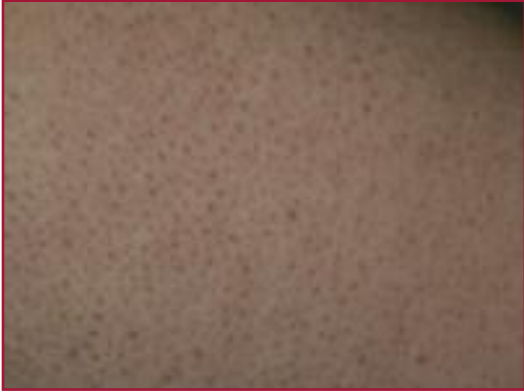
Joseph M. Lam, MD, is a pediatrician with fellowship training in pediatric dermatology who practices in Vancouver, BC

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## Case 7

# *Rough Papules*



A 14-year-old male presents with small, red or tan, rough papules distributed evenly over his upper arms and cheeks.

### *What is your diagnosis?*

- a. Pustular acne
- b. Keratosis pilaris
- c. Rosacea
- d. Atopic dermatitis

### *Answer*

Keratosis pilaris (KP) (**answer b**) is a very common, benign skin condition that is characterized by small (1 to 2 mm), rough, grouped follicular papules that most frequently localize to the posterolateral aspects of the upper arms and anterior thighs. KP can arise in childhood and is known to have an increased incidence during adolescence. For many individuals, KP tends to subside during adulthood.

No treatment is required for this condition, however, application of 12% ammonium lactate lotion, urea cream (10 to 40%), or salicylic acid lotion 6% may help to reduce roughness.

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**Case 8**

## *Hyperpigmented Chest Papules*

An 8-year-old boy presents with hyperpigmented papules over his mid chest area. These have been present for the last few years and they are not pruritic.

### *What is your diagnosis?*

- a. Eruptive vellus hair cysts
- b. Molluscum contagiosum
- c. Flat warts
- d. Scabetic nodules
- e. Darier's disease

### *Answer*

Eruptive vellus hair cysts (**answer a**) are seen as 1mm to 3mm, skin-colored to hyperpigmented (blue to brown) follicular papules, most commonly on the anterior chest. Lesions usually are not grouped and tend to have a smooth surface with a round or dome-like shape. Vellus hair cysts have less commonly been described on the upper and lower extremities, face, neck, abdomen, axillae, posterior trunk, and/or buttocks. They generally occur in children between 4 to 18 years of age. Vellus hair cysts usually resolve spontaneously over months to years. Patients desiring therapy can be treated with incision of individual cysts and expression of their contents, followed by gentle curettage, light electrodesiccation, therapy with topical vitamin A derivatives, lactic acid (12% lotion), or laser.

Molluscum contagiosum is a common cutaneous viral infection in children characterized by singular or multiple skin-coloured papules with central umbilications.

Flat warts, or verrucae plana, occur primarily on the face, neck, arms, and legs. They are usually seen



as smooth, flesh-colored to slightly pink or brown, flat-topped papules measuring 2 to 5 mm in diameter.

Scabetic nodules are red-brown nodules and represent a hypersensitivity response from the host. They occur most commonly on the trunk, axillary regions, and genitalia, and are seen primarily in infants. Although they eventually resolve, scabies nodules may be present for several months.

Darier's disease most commonly first manifests between 8 and 15-years-of-age as flesh-coloured papules that become covered with a yellow waxy scaling crust that can be malodorous. They present in the so-called "seborrheic" distribution, along the forehead, temples, nasolabial folds, scalp, upper chest and back.

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## Case x

# Swelling in Stomach

This gentleman was noticed to have this swelling in his yearly physical examination. It is asymptomatic and is easily reducible.

### *What is your diagnosis?*

- a. Umbilical hernia
- b. Para-umbilical hernia
- c. Lipoma
- d. Sebaceous cyst
- e. Omphalocele

### *Answer*

The answer is Para-umbilical hernia (**answer b**). In adults, most hernias in the umbilical region occur above or below a patient's umbilicus, through a weak place in the linea alba, rather than directly through the umbilicus itself. In Africa, a few of these hernias may be true umbilical ones, which may be so huge that they can accommodate a pregnant uterus.

The typical patient is an obese multiparous woman, with a large multilocular hernia in the upper part of her umbilicus. Its margins are firm, so obstruction and strangulation, particularly Richter type strangulations of the large gut, are common.

If a para-umbilical hernia is small, you should be able to repair it quite easily. Repairing a large one is difficult, because the viscera in the sac stick to its wall, and in freeing them you may damage the gut.

An umbilical hernia is a congenital malformation, especially common in infants of African descent, and more frequent in boys. It is important to distinguish this type of hernia from a Para-umbilical hernia, which occurs in adults and involves a



defect in the midline near to, but not through the umbilicus, and from an omphalocele.

An omphalocele is a congenital malformation in which variable amounts of abdominal contents protrude into the base of the umbilical cord. As the fetus grows during pregnancy, the intestines grow and grow longer and project from the abdomen into the umbilical cord. This growth takes place from the sixth to the tenth week of pregnancy. Normally, the intestines return rapidly into the abdomen by the eleventh week of pregnancy. If this fails to happen, an omphalocele is present. It is important to stress to the parents that they did not do anything to cause the condition. However, more than half of all infants born with an omphalocele may have other birth defects, some of which may be serious.

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.