VIA MEDICA

QUIZ

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Scalp lesion in a newborn: a quiz

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A healthy newborn female infant born vaginally on the term was consulted due to a 4 mm lesion on the scalp that has been noticed just after the delivery (Fig. 1).

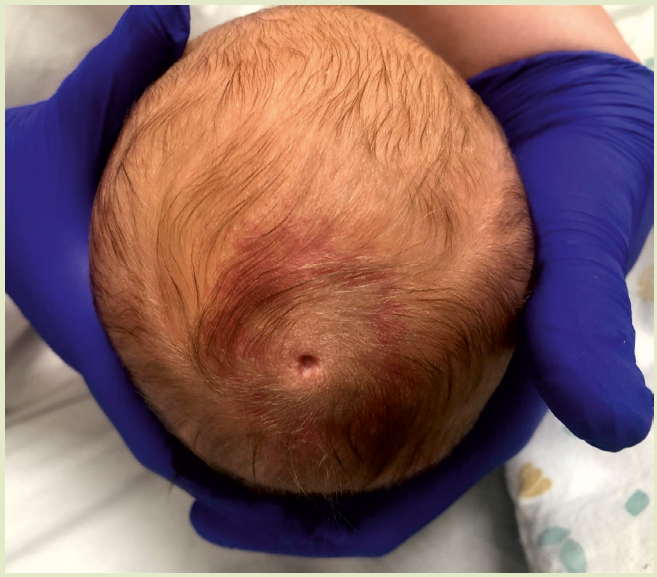

Figure 1. Clinical presentation: lesion on the scalp

Which diagnosis is most likely in our patient?

— Nevus sebaceous
— Labour-related trauma
— Nevus epidermalis
— Neuroblastoma
— Aplasia cutis congenita

See next page for answer.

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ANSWER: APLASIA CUTIS CONGENITA

Aplasia cutis congenita (ACC) is a rare congenital skin disorder. It was first described in 1767 by Cordon but the first lesion on the scalp was d escribed later, in 1826, by Campbell [1]. Aplasia cutis congenita is characterized by a focal or extensive absence of the epidermis, dermis, and occasionally subcutaneous tissue, bone or dura [2, 3]. The disease most commonly occurs on the vertex of the scalp and less often on the trunk or limbs [4]. The condition may be either an isolated congenital defect or less frequently can be associated with defects of the eyes, limbs, gastrointestinal system, genitourinary system or central nervous system [5, 6]. Aplasia cutis congenita has been also reported within a spectrum of genetic syndromes such as Adams-Oliver syndrome, Bart syndrome, and Setleis syndrome [2]. The incidence of ACC is 1 to 3 out of 10 000 newborns with no significant gender or cultural predilection [7, 8].

The aetiology of ACC is probably multifactorial but the exact mechanism is still unknown [3]. Possible factors involved in ACC pathogenesis include chromosomal abnormalities, trauma, amniotic defects, intrauterine vascular ischaemia or infections (i.e. varicella) as well as exposure to teratogens during the pregnancy period (methimazole/carbimazole), angiotensin-converting enzyme inhibitors, misoprostol, cocaine, benzodiazepines, valproic acid, methotrexate) [1, 3, 9, 10]. Aplasia cutis congenita can also be associated with vanishing twin syndrome or epidermolysis bullosa [4, 11].

The pathogenesis of ACC remains unclear. The main pathophysiological hypothesis suggests disruption of the overlying skin between 10–15 weeks of gestation when rapid brain growth occurs along with hair direction and patterning [3].

In 1986 Frieden [5] classified ACC into nine groups taking into consideration the number and the location of the lesions, the presence or the absence of associated malformations and the mode of inheritance.

Group 1: Scalp ACC without multiple anomalies
Group 2: Scalp ACC with limb abnormalities
Group 3: Scalp ACC with epidermal and organoid nevi
Group 4: ACC overlying congenital malformations
Group 5: ACC with associated *fetus papyraceus* or placental infarct
Group 6: ACC with *epidermolysis bullosa*
Group 7: ACC localized to extremities without blistering
Group 8: ACC due to specific teratogens
Group 9: ACC associated with malformation syndromes

Although the diagnosis of ACC is based mainly on clinical presentation, the manifestation of the condition can be very heterogeneous. Typical lesions are small, superficial, noninflammatory, well-circumscribed, and vary in size from a few millimetres to 10 centimetres or even larger. The lesions can have different shapes, and various configurations (circular, oval, linear or stellate) and can be either membranous with a membrane-like surface, round or oval shape or larger non-membranous with irregular shape [1]. Dermoscopy can be useful in clinical diagnostics, including differentiation with a sebaceous nevus. The classic dermoscopic finding of ACC is a "hair collar sign" with hair shafts arranged radially around the lesion, forming a ring of hairs seen through the translucent epidermis [12]. Larger, ulcerative lesions can be associated with underlying morphologic abnormalities like skull defects, cerebrovascular anomalies, or neurological malformations and therefore require further diagnostics [2]. Imaging methods, such as an ultrasound examination and magnetic resonance imaging are useful in the assessment of ACC extent and possible associated anomalies. Similarly, they are crucial for rapid diagnosis of life-threatening complications of ACC such as sagittal sinus haemorrhage, secondary local infections, meningitis and sagittal sinus thrombosis [3, 13].

Managing patients with ACC depends on the size of the lesion, location and presence of associated anomalies. Small lesions on the scalp usually heal with secondary intention within a few months with an atrophic, hairless scar and require only routine wound care [2]. This conservative management includes daily wound cleansing, antiseptic dressing with bacitracin, silver sulfadiazine, or betadine and additional use of parenteral antibiotics in case of systemic signs of infection. Conservative management is introduced mainly in lesions smaller than 4 cm [13, 14]. Larger lesions or ones with an underlying skull defect require early surgical treatment to avoid possible complications, which include haemorrhage, venous thrombosis, and infection [2]. Most performed surgical techniques are primary wound closure, flap repair techniques like local or pedicled scalp flaps with or without primary closure, full-thickness or split-thickness skin grafts for soft tissue coverage and cranial vault reconstruction using bone grafts [3, 15].

Conflict of interest

The authors declare that they have no conflict of interest.

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