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Dermato-venereological Quiz

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This two-month-old baby girl was found to have a patch of alopecia at two days of age. The pregnancy was normal and she was born at full term via normal spontaneous delivery. There was no history of trauma at birth. On examination, apart from the 2x1 cm bald patch near the vertex, there were no other abnormalities. There was no significant family history.

Questions

1. What is the diagnosis?
2. What are the differential diagnoses?
3. What are the possible complications?
4. What is the prognosis and management of this condition?

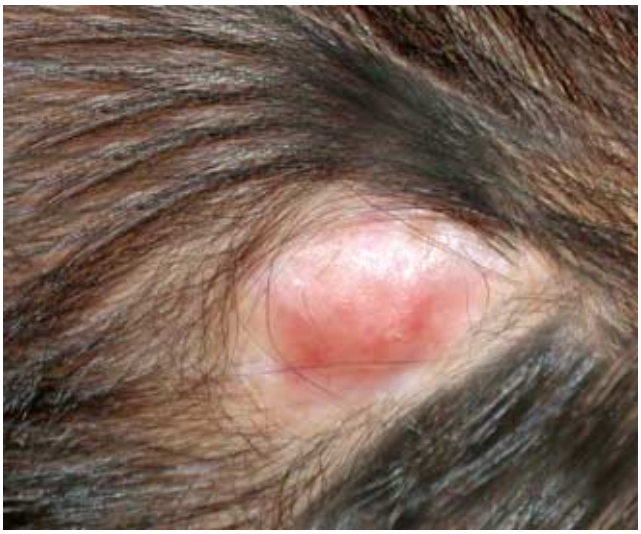

Figure 1.

(Answers on page 122)

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Answers to Dermato-venereological Quiz on page 107

1. The diagnosis is aplasia cutis congenita (ACC). It is due the failure of fusion of skin and occurs in 1 per 3000 births. Histologically, there is absence of epidermis and in most cases, the dermis. The subcutaneous fat may also be absent. ACC may be inherited in an autosomal dominant trait in some cases. Lesions most commonly occur on the scalp near the hair whorl but can also be found on the proximal extremities, trunk or face. They are single in about 70% cases and the size may vary from 0.5 to 10 cm in diameter, with variable depth and shape. The lesions are sharply demarcated and non-inflammatory in appearance. Lesions may be ulcerated or healed with a superficial membrane and occasionally bulla formation may be found. There may also be adjacent hypertrichosis.

2. The differential diagnoses include birth trauma secondary to forceps, vacuum extraction or fetal scalp electrodes, sebaceous naevus and focal dermal hypoplasia.

3. Lesions may extend to the scalp, dura or meninges. Such lesions may lead to infection, haemorrhage, sagittal sinus thrombosis or meningitis. Bone involvement may be present in a third of cases. ACC may be associated with cleft lip or palate, syndactyly, absence of digits, and congenital heart disease.

4. The majority of lesions will heal spontaneously from the edges to form a thin hairless scar although hypertrophic scarring occasionally occurs. The prognosis is good if secondary infection and trauma are avoided. Skin grafting may be required for large defects to prevent haemorrhage or infection of the sagittal sinus. Large cranial defects may need bone grafts.