Port-wine stain associated with membranous aplasia cutis congenita and hair collar sign

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DESCRIPTION

A newborn of around 2 weeks was referred by a paediatrician for the evaluation of a bald area, surrounded by a large reddish patch on the scalp. Skin examination revealed a flat pinkish to reddish patch (figure 1A) with a round, atrophic area of alopecia, 20 mm in diameter, with a membranous surface in the centre, accompanied by a rim of terminal hairs (figure 1B). Pregnancy and the delivery had been unremarkable and the baby was otherwise healthy. There was no deepening of the colour of the red patch with vigorous activities of the baby like crying or breath holding. Dermoscopy of the central part showed a reddish background, thin linear vessels and remarkably few hair bulbs (figure 1C). No skull bone and brain defects were found. Dermoscopy of red surrounding areas showed red, rounded, globular vessels representing the superficial or papillary form of port-wine stain (PWS), disclosing vertically oriented capillaries (figure 1D). The lesion was diagnosed clinically as membranous aplasia cutis (MAC) with a hair collar and naevus flammeus supported by dermoscopic findings.

Aplasia cutis congenita (ACC) is a congenital defect of the skin characterised by localised absence of the epidermis, dermis and, at times, underlying structures such as bone or dura.[1] It generally occurs on the scalp. The existence of maximum tensile force during rapid brain growth in that region is the possible explanation for the location of ACC on the vertex. The various clinical presentations may be bullous, fissure-like ulcers with a granulating base, erosions, atrophic macules or scars.[1 2] MAC congenita is a cystic variant covered with a membranous or glistening surface.[1]

'Hair collar' sign is suggested to have a close association with neuroectodermal defects. A failure of the normal closure of the cranial neural tube may affect the fetal skin development, including melanoblast migration and capillary network formation giving rise to association with dermal melanocytosis and the other with naevus flammeus.[1 3]

The skin and the nervous system are derived from the ectoderm. This embryological association may explain the cutaneous abnormalities often found overlying neural tube defects. The 'hair collar sign', a ring of dark long hair encircling a congenital scalp lesion, is one such marker.

PWS needs to differentiate from the salmon patch in the index case. PWSs present at birth as sharply demarcated red macules or patches. The lesions' colour changes from dark red to violaceous as age advances. PWSs can occur anywhere on the body;

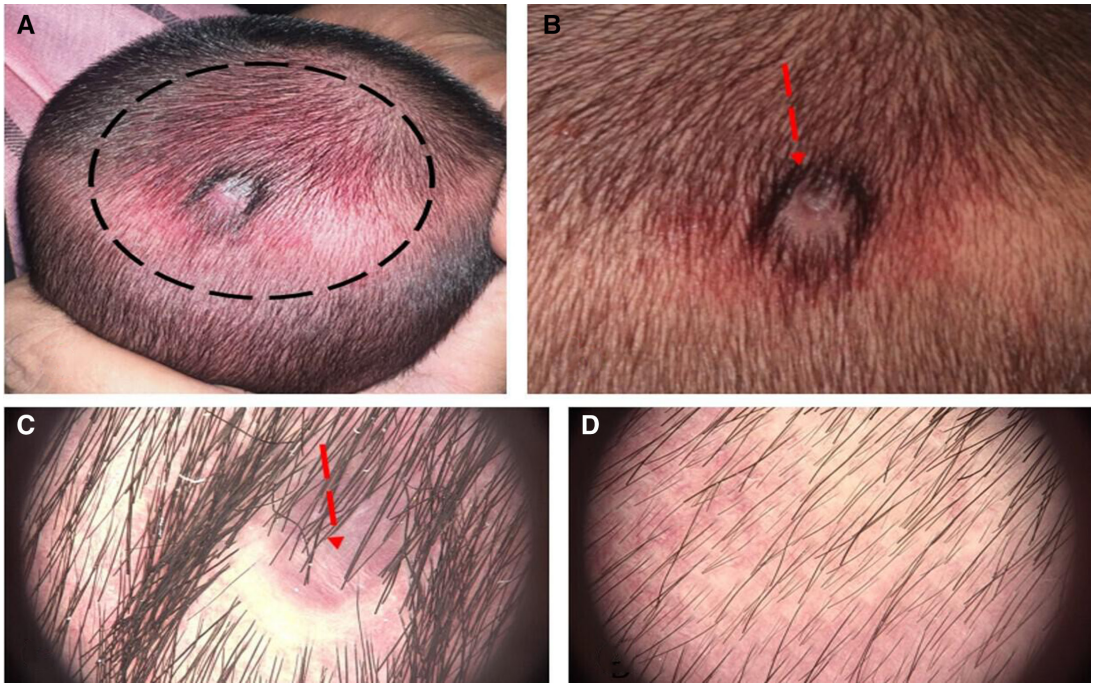

Figure 1 Superior/posterior location of scalp showing port wine stain (A) and membranous aplasia cutis congenita with 'hair collar' sign (B) in the vertex and dermoscopic findings (C, D).

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the common site being the face. They persist throughout life growing with the child. Salmon patches are scarlet to pink and flat, can be totally blanched and usually deepen in colour with vigorous activity like crying, straining with defecation, breath holding and with changes in ambient temperature. The common site of lesions being on the nape, followed by the glabella and eyelids. In spite of their midline location, they are not associated with spinal dysraphism except those in the sacral area. Salmon patches tend to resolve or significantly regress with time.[4 5]

In conclusion, the changes in the index cases, as well as the hair collar sign, may suggest a complex hamartomatous nature of MAC. Red macule, not becoming prominent on crying or breath holding of the baby, and location on the scalp suggest the lesion is PWS rather than a salmon patch. The presence of a vascular stain and hair collar sign with MAC or a congenital scalp nodule should increase suspicion of an associated cranial dysraphism.

Learning points

- A rim of hypertrichosis, 'hair collar' sign, is proposed to have a close association with neuroectodermal defects.
- Membranous aplasia cutis is a form fruste of a neural tube defect.
- The presence of a vascular stain and hair collar sign with or without a congenital scalp defect should increase suspicion of an associated cranial dysraphism.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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