Occult craniocervical dysraphism and skin markers

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DESCRIPTION
Neural tissue and skin are of common ectodermal origin and therefore anomalies occurring during early embryogenesis may lead to combined neuroskin malformations. Neural tube closure occurs during primary neurulation starting from the equivalent of the craniocervical junction and proceeding ziplike in both cephalic and caudal directions. Therefore, any defects of the process may reflect spinal dysraphism with, possibly, combined congenital skin lesions.

Occult spinal dysraphism (not exposed but skin-covered neural tissue), in the vast majority of cases, is localised in the lumbosacral area and less commonly in the cervicothoracic area. This report describes an atypical case of occult craniocervical dysraphism with combined skin lesions/markers.

Routine postnatal examination of a term newborn girl revealed a salmon patch (vascular malformation of capillaries) on the occipital and upper cervical areas, a subcutaneous soft but fixed mass (1.3×1.2×1.0 cm) on the midline at the level of the craniocervical junction, and hair tufts above and below the mass along the midline (figure 1). The differential diagnosis of meningocoele/myelocele, lipoma, dermal cyst, fibroma, or hamartoma was made. Brain and spine MRI revealed a complex anomaly of the craniocervical junction with a fluid density tract (figure 2) extending from the cisterna magna, and a cerebrospinal fluid space, through a bony defect, to a small subcutaneous cyst (figure 3), providing evidence of an occult occipital meningocoele. There was no herniation of the cerebellum or intracranial contents, and no abnormalities of the lower spine.

Figure 1 Salmon patch, subcutaneous soft mass and hair tufts along the midline of occipital and craniocervical areas.

Figure 2 Fluid density tract (T2 TSE sequence, sagittal view, arrow) protruding through the bony defect.

Figure 3 Subcutaneous cyst/occult occipital meningocoele (T2 TSE sequence, axial view, arrow).
Learning points

▸ Craniocervical dysraphism represents an anomaly with its origin in early embryogenesis (week 3).
▸ Neural tissue and skin are of common ectodermal origin and may therefore present combined neuro-skin malformations.
▸ Occult spinal dysraphism with skin markers may be atypically located in the craniocervical area.

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REFERENCES