Hair collar sign

The “hair collar sign”, a designation introduced by Commens et al. in 1989, is characterized by the presence of a ring of terminal hairs around any cutaneous lesion. It has been described as a relatively specific marker of neuroectodermal defects.

Infants with Hair collar sign (HCS) and hair tuft of the scalp (HTS) are at high risk for underlying neurovascular anomalies. Magnetic resonance imaging scans should be performed in order to refer the infant to the appropriate specialist for management [1].

Kuemmet et al. investigated the risk of cranial central nervous system dysraphism in children presenting with aplasia cutis congenita of the head, who presented between 1/1/2000 and 6/15/2016. Inclusion criteria were subjects with aplasia cutis congenita of the head who received CT or MR imaging of the head.

They identified a total of 69 subjects with aplasia cutis congenita affecting the head and who received imaging. The most common location of the aplasia cutis congenita lesion was the vertex scalp (49.3%). The hair collar sign was present in 27.5% of patients. Twelve of 69 patients (17.4%) demonstrated abnormalities of the bone, vasculature, or brain on head imaging. Only one patient had a diagnosis of encephalocele that required neurosurgical intervention. There was a statistical association between the hair collar sign and the presence of abnormal imaging findings (P = .029), with a negative predictive value of 89.4%.

The incidence of central nervous system dysraphism in patients with aplasia cutis congenita of the head appears below, and it may not be necessary to image the head of each child presenting with this skin lesion. The hair collar sign may be a marker of underlying defects [2].

Takayama et al. reported two cases of aplasia cutis congenita associated with hair collar signs and hemangioma simplex in their parietal regions. A hair collar sign and a hemangioma are known to suggest the possibility of underlying neural tube defects. However, no obvious bone defects or heterotopic neural tissue were observed in the imaging and histopathological examinations. Nevertheless, some pathological observations similar in both cases suggested abnormalities in the process of ectodermal fusion. A flat epidermis and a lack of appendages were recognized. Both cases also exhibited the presence of melanocytes in a portion of the superficial dermal layer. An increase in the number of macrophages was observed in the dermal area with neither elastic fibers nor normal

collagen fibers. The peripheral hair follicles grew horizontally [3].

Hair-Collar-and-Tuft-Sign Associated with an Atretic Cephalocele and a Persistent Primitive Falcine Sinus [4].

References

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