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Concerning Simpson and Goodman’s Paper:

Scalp Hamartoma or Atretic Cephalocele?

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Dear Sir,

We read with interest the article ‘Scalp hamartoma in identical twins’ written by Simpson et al. [1]. The fact of this occurrence in identical twins is very interesting indeed, as is the discussion on the genetic basis of this presentation. Nevertheless, after having finished the reading of this report, we were left with the doubt of what type of lesion the authors are referring to. We understood that they are mentioning atretic cephalocele. Accordingly, we would like to make some remarks, with your kind permission.

(1) In their introduction, the authors affirm that the present literature on subscalp tumors is confusing in regard to nomenclature. Nevertheless, there are several important studies dealing with tumors of the cranial vault in general [2, 3] and with cephalo-celes in particular [4-7] that may help to clarify both the terminology and the concept of subscalp neoplasms. Certainly, the nature of scalp and cranial tumors in children may remain enigmatic until studied microscopically [2]. However, a combined clinicopatho-logical approach to these scalp lesions seems to be very important in establishing the diagnosis [2, 3]. The term atretic cephalocele refers to a skin-covered subscalp lesion that consists of nonspecific fibrous tissue, devoid of cutaneous adnexae, and that sometimes contains a prominent vascular net and neu-rogliar rests [7, 8]. They might represent spontaneous arrest in the development of a true encephalomeningocele. In cephalocele, there is a communication between the subscalp nodule and the central cavity [4, 8]. Thus, a main point for the distinction between the cephalocele and glial (or menin-geal) heterotopia seems to be the presence or absence of a bone defect [4, 7]. Perhaps, the difference in terminology between scalp ha-martomata and atretic cephaloceles depends more on the histological than on the clinical approach to these lesions.

In the description of the case report, it would have been convenient to mention the radiological investigations they performed to rule out intracranial involvement. The study of a midline lesion in a child’s scalp must include a detailed neuroradiological study, either by computed tomography scan or by magnetic resonance imaging [8]. Some of these benign scalp tumors may have a significant vascular component [5], a fact that is relevant at the time of surgical excision, or they may associate with important cerebral malformations [6, 8]. It is the underlying brain involvement that determines the prognosis and not the small cephalocele.

We agree with the authors in that the main indications for excision are obtaining a histological diagnosis and the relief of the pain that the children with these cysts complain of, and not only parental anxiety. In our opinion it is unwise to leave these lesions, as other scalp tumors, untreated if their nature is not clearly established from complementary investigations [9].

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Richard K. Simpson, Jr. J. Clay Goodman
Reply

Dear Sir,

Dr. Goodman and I would like to provide a brief comment to the letter by Dr. J.F. Martínez-Lage regarding our paper entitled 'Scalp Hamartoma in Identical Twins', recently published in *Pediatric Neurosurgery* [1993;19:89-92].

We wish to thank Dr. Martinez-Lage for his thoughtful comments about our paper. We regard cephaloceles and scalp hamartomas as extremes of a continuum of lesions with impaired neural tube closure being the unifying principle. As Dr. Martinez-Lage suggests, the presence or absence of neuroradiologically demonstrable skull defects may serve as a convenient clinical means of distinguishing these lesions. We mentioned that our radiographic studies revealed no other extracranial or intracranial pathology. The emphasis of our paper was the occurrence of such lesions in identical twin males. Although shared intrauterine environmental challenges to the fetus may be the etiology, a genetic mechanism including sex-linked transmission is also a distinct possibility.

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