Steroid sulfatase (STS) enzyme (EC 3.1.6.2) is present in the microsomal fraction of the cell and is capable of hydrolyzing sulfated sterols (Hobkirk, 1985; Dibbelt and Kuss, 1991). STS deficiency is the biochemical defect in X-linked ichthyosis (XLI), a disease that clinically presents dark, adhesive, and regular scales of skin (Shapiro and Weiss, 1978). STS assay or FISH analyzes allow diagnosis of XLI and identify XLI-carriers (Baden et al, 1980; Epstien and Lевенштейн, 1981; Okano et al, 1985; Herrmann et al, 1987; Matsumoto et al, 1990; Aviram-Goldring et al, 2000). STS enzyme is encoded by the STS gene on the short arm of the X-chromosome ( Muller et al, 1981). Most STS enzyme deficient patients present large deletions of the entire STS gene and flanking sequences (Bonifas et al, 1987; Shapiro et al, 1989; Cuevas-Covarrubias et al, 1997; Aviram-Goldring et al, 2000). Only a few point mutations or partial deletions have been identified (Bonifas et al, 1987; Ballabio et al, 1989; Shapiro et al, 1989; Basler et al, 1992; Nomura et al, 1995; Alperin and Shapiro, 1997; Morita et al, 1997; Aviram-Goldring et al, 2000; Oyama et al, 2000; Sugawara et al, 2000; Valdes et al, 2000). In this study, we describe an XLI patient with the first 3 bp intragenic deletion of the STS gene causing XLI and demonstrate the maternal transmission of this molecular defect.

MATERIALS AND METHODS

Patient The patient was referred as having ichthyosis to the General Hospital of Mexico. He was the only child of the family. The patient and his mother were informed about the characteristics of the study and they agreed to participate. Protocol was evaluated and approved by the Ethics Committee. The patient showed a moderate degree of

REFERENCES


Expression of Basal-Cell Adhesion Molecule (B-CAM) in Human Epidermis

To the Editor:

I read with interest the recent article by Schön et al on the expression of basal-cell adhesion molecule (B-CAM) in human skin (Schön et al, 2000) documenting association of B-CAM expression with activated states of keratinocytes. The authors provide convincing evidence that, besides its known function as a laminin receptor, B-CAM may be involved in cell-cell interaction or migration.

In this context I would like to bring to your attention data regarding B-CAM expression in human skin that might not have been accessible to the authors at the time their manuscript was submitted. We have been analyzing B-CAM expression in normal and diseased human skin utilizing the monoclonal antibody G253 that has also been employed in the paper by Schön et al. Contrary to their observation, but in line with previous findings (Garin-Chesa et al, 1994), the majority of specimen from normal human epidermis exhibited B-CAM positive suprabasal keratinocytes. In contrast, B-CAM expression in fetal epidermis was restricted to basal keratinocytes. The outer root sheath of hair follicles in both fetal as well as adult human skin regularly exhibited a B-CAM positive phenotype. The level of B-CAM expression was higher in psoriasis and contact dermatitis. We interpret our observations as evidence for B-CAM being a keratinocyte differentiation marker (Bernemann et al, 2000). This notion is supported by the effects of Ca2+ levels on the B-CAM phenotype of keratinocytes as described by Schön et al.

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REFERENCES


Maternal Transmission of the 3 bp Deletion within Exon 7 of the STS Gene in Steroid Sulfatase Deficiency

To the Editor:

Steroid sulfatase (STS) enzyme (EC 3.1.6.2) is present in the microsomal fraction of the cell and is capable of hydrolyzing sulfated sterols (Hobkirk, 1985; Dibbelt and Kuss, 1991). STS deficiency is the biochemical defect in X-linked ichthyosis (XLI), a disease that clinically presents dark, adhesive, and regular scales of skin (Shapiro and Weiss, 1978). STS assay or FISH analyzes allow diagnosis of XLI and identify XLI-carriers (Baden et al, 1980; Epstien and Lевенштейн, 1981; Okano et al, 1985; Herrmann et al, 1987; Matsumoto et al, 1990; Aviram-Goldring et al, 2000). STS enzyme is encoded by the STS gene on the short arm of the X-chromosome ( Muller et al, 1981). Most STS enzyme deficient