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Introduction

AS DERMATOLOGISTS, one of the most important things we may do is to detect subtle cutaneous clues that aid in the diagnosis or recognition of significant underlying disease. Pediatric patients are often referred to dermatologists for evaluations of unusual "birthmarks" or atypical rashes. In most cases, these are reflections of benign processes that require simple reassurance or straightforward treatment. However, in certain cases, subtle cutaneous signs can herald the presence or onset of significant underlying disease. Dermatologists should be aware of these cutaneous markers, the clinical characteristics that predict higher risk, and the appropriate diagnostic tests and monitoring required to care for these patients.

This issue of *Seminars in Cutaneous Medicine and Surgery* highlights several dermatologic findings that have been associated with abnormalities in the nervous system. This issue is by no means comprehensive, and there are other important disorders including neurofibromatosis and tuberous sclerosis, which are not reviewed here, but have been described extensively elsewhere in the literature. The authors in this issue discuss several pediatric conditions that have potential systemic, including neurologic implications.

These disorders are characterized by a wide array of physical findings. Skin lesions may range from remnants of the defective fusion of developmental units to tumorous growths and malformations to manifestations of genetic mosaicism. The first article reviews the cutaneous manifestations of underlying neural tube defects, and describes a spectrum of findings from obvious developmental defects, as in the case of meningomyelocele and spina bifida, to more subtle lesions such as aplasia cutis. Some of the associated skin manifestations discussed in this issue of *Seminars* are considered "birthmarks" because of their appearance at birth or early in life. In general, the term "birthmark" has a benign, reassuring connotation; it is true that most congenital nevi, vascular stains, and pigmentary anomalies are not associated with underlying disorders. However, there are certain subsets of "birthmarks" that should prompt further evaluation. Port-wine stains and congenital nevi are relatively common "birthmarks" that typically attract little attention and are only rarely associated with neurologic sequelae. This issue includes articles about Sturge Weber syndrome and neurocutaneous melanosis that describe clinical characteristics that help to distinguish patients at risk for neurocutaneous disease from those with less worri-

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some lesions. Benign growths such as epidermal hamartomas and hemangiomas have been associated with several extracutaneous findings and are each discussed separately in the following articles. The location, extent, and morphologic characteristics of these lesions have been recognized as important clues to extracutaneous involvement. The persistence of prominent vascular markings and atrophy in cutis marmorata telangiectatica congenita represents a less common entity that is also associated with a number of unusual anomalies. The final article discusses incontinentia pigmenti, in which cutaneous signs may manifest as inflammatory pustules, verrucous lesions, or pigmentary changes.

The link between cutaneous markers and neurologic disease is still poorly understood in many disorders. However, the growing understanding of genetics and the molecular mechanisms underlying tissue development and function has led to several theories of pathogenesis in these disorders. Several candidate genes have been implicated in variants of epidermal nevus syndrome and genetic mosaicism is believed to explain the observed phenotypic patterns. Finally, the recent elucidation of the *NF-KB* pathway has not only expanded our understanding of disorders (ie, incontinentia pigmenti) in which components of this pathway are defective, but also has broadened our understanding of normal physiology. While many of the associations described in this issue of *Seminars* may still not be fully explained, the mysteries may be unraveled as our scientific knowledge continues to grow.

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Guest Editor