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Generalized Atrophic Dells in a Newborn

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Infantile myofibromatosis (IM) is a nonmetastasizing locally invasive neoplasm. The behavior of the tumor is more hamartomatous than tumoral, and it is unclear whether the cell of origin is a fibroblast or a smooth muscle myocyte. Lesions typically present during infancy and range in size from a few millimeters to several centimeters. We present an unusual case of a patient with an atrophic variant of IM.

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Case Report

A 3-month-old boy was referred for evaluation of 10 to 15 scarred lesions on his back, sacrum, trunk, and extremities. The patient's mother noted these lesions at birth, and the lesions have neither changed nor spontaneously bled since birth. The patient was born at 39 weeks by vaginal delivery to a healthy 32-year-old primigravida. His weight at birth was 7 lb, 7 oz, and his Apgar score was 9/9 at 1 and 5 minutes. The pregnancy was uncomplicated, and routine prenatal care revealed no comorbid conditions throughout gestation. The patient's mother noted a personal history of varicella at 5 years old, which spontaneously resolved without sequelae. There was no family history of syndromic conditions.

Physical examination revealed an alert infant who appeared to be well-developed and well-nourished. There was no evidence of dysmorphism. Cutaneous examination revealed lesions with 2 morphologies: atrophic dells with a bluish or telangiectatic center (Figure 1), and firm bluish-red papules. These discrete papular lesions were found on the infant's left anterior chest (Figure 2) and hypogastrium and measured 3 to 4 mm in diameter. The nails and mucosal membranes were unaffected. There was no associated lymphadenopathy or hepatosplenomegaly. Diagnostic skin biopsies were obtained from the atrophic dells and bluish-red papules.

Histopathology—Histologic sections viewed at low and high magnification revealed an unaffected epidermis. Both the atrophic dells and papules revealed a proliferation of spindle cells arranged in whorls and bundles within the dermis (Figure 3). The spindle cells had round or oval nuclei and elongated cytoplasmic processes. There were low numbers of mitoses (4 of 10 high-power fields). Atypical mitoses were not seen. A smooth muscle actin stain was positive. A trichrome-stained biopsy section revealed small bundles of collagen in between tumor bundles. An S100 protein stain was negative.

Clinical Course—Results of magnetic resonance imaging (MRI) of the brain, chest, abdomen, and pelvis revealed no additional lesions. No new lesions developed and the majority of those lesions present resolved over several months. Papules noted on initial examination evolved into atrophic dells before disappearing completely.

Comment

Infantile myofibromatosis (IM), first described by Chung and Enzinger[1] in 1981, is a nonmetastasizing locally invasive neoplasm. IM is considered the most common fibrous proliferation in children.[2] The behavior of the tumor is more hamartomatous than tumoral, favoring slow local growth over metastasis. It is unclear whether the cell of origin is a fibroblast or a smooth muscle myocyte. Lesions typically present during infancy and range in size from a few millimeters to several centimeters. Chung and Enzinger[1] found that 54 (88%) of 61 patients with myofibromatosis exhibited lesions before the age of 2 years, and only 14 patients (26%) had visceral involvement. The scarring or atrophic presentation described in our patient, though exceedingly rare, also has been reported in the literature.[3]

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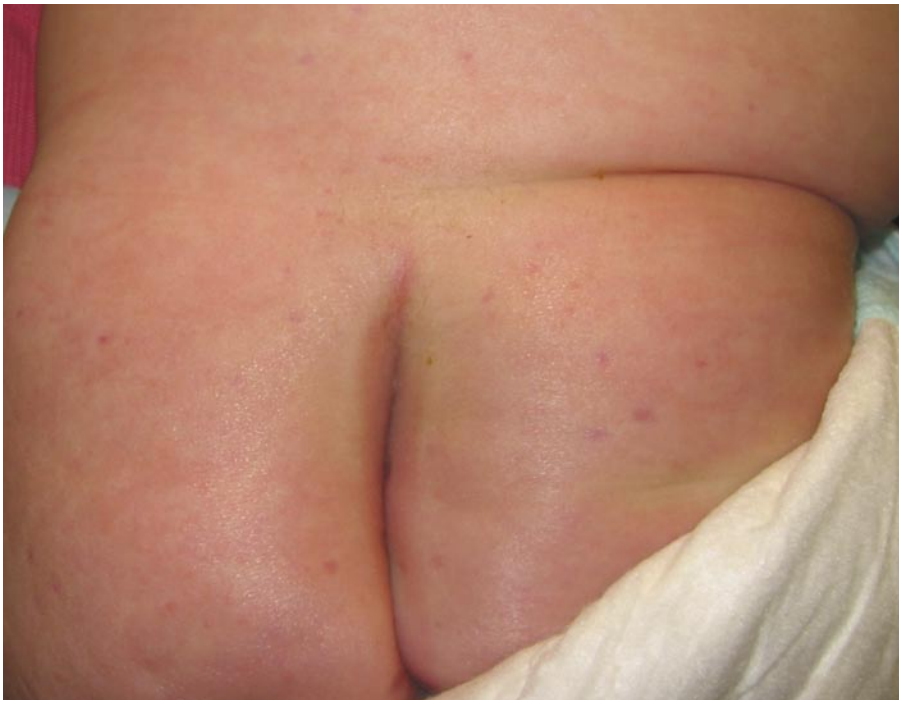

Figure 1. Atrophic dells on the infant's trunk and extremities, predominately along the sacrum.

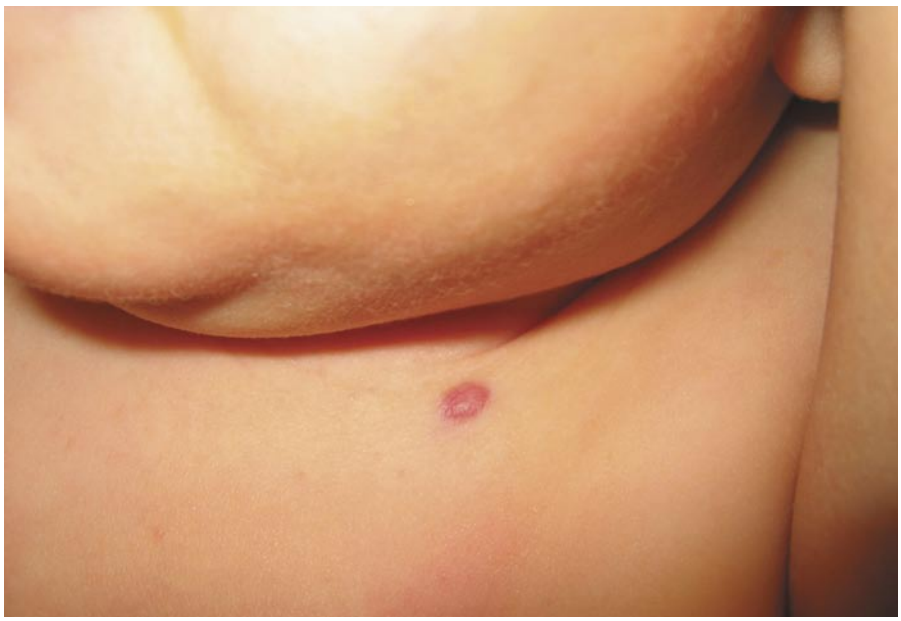

Figure 2. Discrete reddish papules measuring 3 to 4 mm in diameter on the left anterior chest.

Solitary IM occurs more often in infant boys and classically involves the soft tissues of the head and neck.[3] Solitary IM comprises 80% of all myofibromatoses. Multicentric IM is more common in infant girls, involves the viscera and bone in 25% to 35% of patients, and portends a worse prognosis.[4] Multicentric IM is subdivided further into types 1 and 2.[5] Type 1 involves the skin, subcutaneous tissue, and muscle, without visceral involvement. Type 2 involves the viscera, and type 2A is the most rapidly fatal variant, with overwhelming lung involvement and subsequent respiratory failure. Most visceral lesions are present at birth and exhibit no apparent site of predilection.[5] Bony lesions, when found, most commonly affect the long bones.[6]

The pathogenesis of IM tenuously has been linked with estrogen levels.[7] The tumor is a proliferation of spindle cells that group together to form curved bundles.[8] These bundles give a whorled or nodular pattern to the tumor. Occasionally, necrosis may be seen in the center of these nodules. Electron microscopy reveals a cell-rich proliferation of both

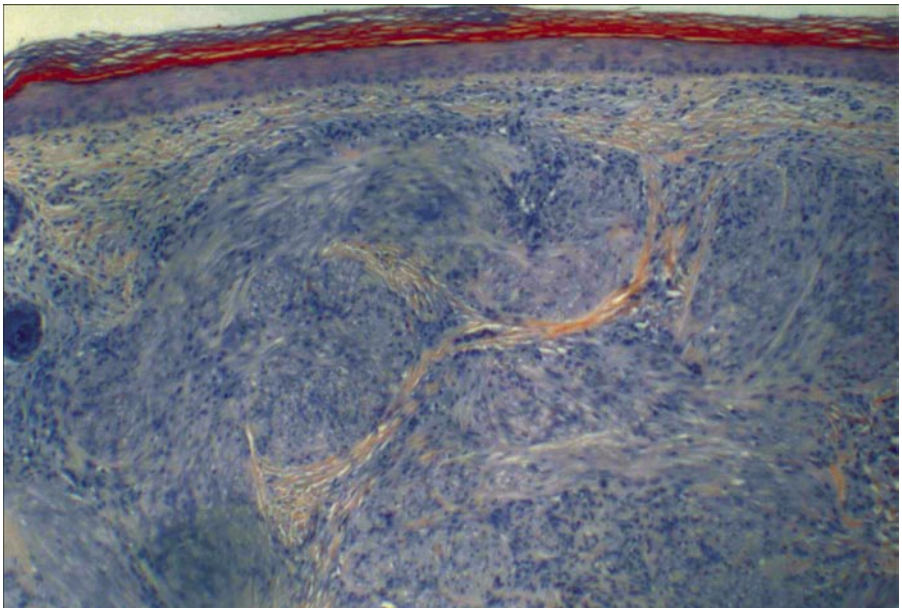

Figure 3. Whorls and bundles of spindle cells characteristic of infantile myofibromatosis (H&E, original magnification ×20).

fibroblasts and myofibroblasts arranged haphazardly.[8] A smooth muscle actin stain typically is positive, supporting the proposed cell of origin. A trichrome will stain the tumor red for muscle. An S100 protein stain should be negative in IM, which is reassuring to the clinician that a neural or melanocytic process is not at play.

The differential diagnosis for IM includes histiocytosis, hemangiomatosis, neurofibromatosis, atrophic dermatofibromas, leukemia cutis, lymphoma cutis, neuroblastoma, and extramedullary hematopoiesis.[9] Juvenile fibromatosis, which tends to be poorly circumscribed, present in slightly older children, and behave more aggressively, also is included in the differential diagnosis. A tissue sample almost always is required to confirm the diagnosis of IM. If there are more than 20 skin lesions present, the clinician is compelled to perform an MRI on the infant to rule out visceral or other systemic involvement. In our experience, a full body MRI is the most cost-effective modality, with the highest sensitivity; however, alternative imaging techniques such as echocardiography and ultrasonography also are found in the literature.[9]

Treatment of IM depends on appropriate staging.[10] Resection of solitary tumors typically is reserved for symptomatic lesions because solitary IM may involute spontaneously. The rate of recurrent lesions following excision ranges from 7% to 10%. For multicentric IM involving viscera, a combination of chemotherapeutic agents has been used with mixed results.[10] Periodic imaging is recommended until the condition has stabilized and no new lesions are found.

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