63

#### CASE MONTH O F THE

#### Diagnosis: Myofibroma

Infantile myofibromatosis is a rare and potentially life-threatening condition that should be considered in children who present with multiple skin nodules, Dr. Ivan D. Camacho reported at the annual pre-AAD meeting of the Society for Pediatric Dermatology.

'To our knowledge, this is the first report of a case of myofibromatosis of infancy confined to the genital area," said Dr. Camacho, a dermatology fellow at the University of Miami.

The myofibroma in this case was diagnosed following a punch biopsy, which demonstrated proliferative, plump, spindle-shaped cells forming interweaving fascicles imbedded in a fibromyxoid stroma. Smooth muscle actin and vimentin stains were positive, and desmin, cytokeratin, and S-100 stains were negative, which is consistent with the diagnosis of myofibroma, he said.

Myofibromas are rare, mesenchymal tumors that can present as a single lesion or in groups. They are firm, skin-colored to violaceous nodules that can involve the skin, subcutaneous tissues, striated muscles, bones, and viscera. Multiple lesions in a newborn or infant are known as infantile myofibromatosis. In most cases, myofibromas occur as solitary lesions that rarely involve the viscera.

Multiple lesions, however, often involve soft tissues, bone, and viscera and are associated with adverse prognosis and outcome, with mortality resulting from airway obstruction, intestinal obstruc-

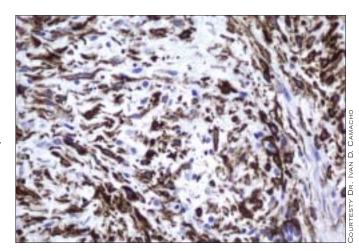
tion, failure to thrive, or infection. In rare cases, the brain, orbit, and testis might be involved.

The lesions should be differentiated from those associated with neurofibromatosis, fibrous hamartomas of infancy, sarcomas, granuloma gluteale infantum, and vascular tumors. Patients with multicentric cases should undergo

systemic evaluation for involvement of the viscera because surgery, chemotherapy, and radiotherapy might be beneficial in those with visceral involvement. Patients without visceral involvement have an excellent prognosis, with the lesions typically regressing spontaneously over 1-2 years, as was the case in this child, Dr. Camacho said.

At follow-up, the patient's lesions were resolving spontaneously and his mother refused further work-up.

-Sharon Worcester



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# INDEX OF ADVERTISERS

Allergan, Inc.	
Juvederm	19
Abbott Laboratories	
Humira	12a-12f
Astellas Pharma US, Inc.	
Protopic	56a-56b
Bayer HealthCare Pharmaceuticals In	
Taz	11-12
Beiersdorf Inc.	
Eucerin	18
Centocor, Inc.	
Remicade	40a-40d, 41-42
CORIA Laboratories, Ltd.	
Salex	7-8
Atralin	23-24
Ellipse Inc.	
JUVIA	55
Galderma Laboratories, L.P.	
Tri-Luma	5-6
Differin	16a-16b
Clobex	24a-24b, 48a-48b
Clindagel	29-30
Metrogel	39-40
Graceway Pharmaceuticals, LLC	
Aldara	22, 43-45
INTENDIS, Inc.	
Finacea	60a-60b
Lexington International, LLC	
HairMax LaserComb	46
Medicis Pharmaceutical Corporation	
Vanos	8a-8b
OrthoNeutrogena	2.4
Retin-A Micro Renova	3-4 37-38
	37-38
PCA Advanced Skin Care Systems	
Hydrating Serum	56
Ranbaxy Laboratories, Inc.	
Halog	21
Ultravate and Lac-Hydrin	51
Eurax	59
sanofi-aventis U.S. LLC	
Xyzal	47-48
Stiefel Laboratories, Inc.	
Olux-E	14-16
MimyX	27-28
Evoclin	52a-52b
Duac	64
Wyeth Pharmaceuticals Inc.	
Enbrel	31-34

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