

Tufted angioma*

Camila Mendes da Silva¹
 Antonio Pedro Mendes Schettini²
 Monica Santos^{3,4}
 Carlos Alberto Rodrigues Chirano⁵

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Dear Editor,

Tufted angioma is a rare vascular tumor, with slow angiomatous proliferation, usually located in the skin and subcutaneous tissue. Its clinical presentation is of a solitary angiomatous tumor, usually located on the neck, upper trunk and extremities. It is more frequent in children, with no sex or racial predilection. The term tufted angioma (TA) is due to the dense agglomerates of endothelial cell lobules and capillaries on histology.¹ This is a case report of a seven-year-old female patient with a painful, rapidly growing lesion over the past 3 months on the right flank. On dermatological examination, a hardened angiomatous tumor is present, with irregular surface, its larger diameter measuring approximately 3cm on the right flank (Figure 1). An incisional biopsy was performed, and histopathology revealed epithelial hyperplasia and proliferation of vascular structures in a glomerular pattern, and a "cannonball" pattern of the vessels in the superficial and deep dermis, diagnostic of vascular neoplasia (Figure 2). Immunohistochemistry revealed focally positive CD34 antibody - against hematopoietic cells and pericytes clone QBEnd10) - and positive CD31 antibody - against adhesion CD31 adhesion molecule PECAM-1 (endothelial cells) (clone JC/70A). Cytokeratins 40, 48, 50 e 50,6kDa (clone AEI/AE3), desmin (muscle cell intermediate filament) (clone D33), protein S-100 (polyclonal clone) and smooth muscle actin (clone 1A4) analysis was negative. In view of the clinical and laboratory findings, the diagnosis of a low-grade vascular proliferation: acquired tufted angioma (angioblastoma of Nakagawa) was made. The le-



FIGURE 1: Indurated angiomatous tumor with irregular surface, larger diameter measuring approximately 3cm, located on the right flank

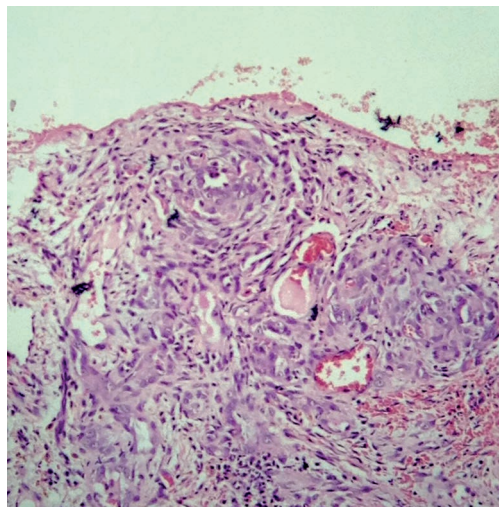


FIGURE 2: Vessels in the dermis with a "cannonball" pattern and capillary structures in a glomeruloid arrangement. Hematoxylin & eosin X100

sion was excised with an ellipse down to the muscular fascia, with a lateral margin of 0.5cm. The patient was reviewed three months after surgery and had good functional outcome despite the hypertrophic scar, for which was prescribed topical gel with cepalin + heparin, t.i.d. In 1949, Nakagawa described a proliferative vascular entity which he named angioblastoma. In 1976, Wilson Jones used for the first time the term tufted angioma to name an acquired vascular proliferation.^{2,3} TA can be congenital or acquired but, in approximately 50% of reports, the lesion appears within the first year of life.¹ Its pathogenesis has not been established; however, the increase in endothelial and vascular growth factors is involved in its angiogenesis, allowing for the development of capillary lobes.³ Trauma can be a contributing factor. It can present in three different clinical patterns: uncomplicated TA (most common type), TA without thrombocytopenia but with chronic coagulopathy and TA complicated by Kasabach-Merritt syndrome with thrombocytopenia.¹ Clinical presentation consists of a macule or papule similar to a Port-wine stain that progresses to an angiomatous tumor, varying

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¹ Private practice - Manaus (AM), Brazil.

² Dermatopathology Service - Fundação Alfredo da Matta (FUAM) - Manaus (AM), Brazil.

³ Teaching and Research Sector - Fundação Alfredo da Matta (FUAM) - Manaus (AM), Brazil.

⁴ Dermatology Department - Universidade do Estado do Amazonas (UEA) - Manaus (AM), Brazil.

⁵ Dermatologic Surgery Service - Fundação Alfredo da Matta (FUAM) - Manaus (AM), Brazil.

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in size from 2 to 10cm, most often found on the upper trunk, cervical area, and proximal aspect of the limbs, besides other locations such as face and oral mucosa.¹ Lesions are usually asymptomatic, but can be very painful and present with hypertrichosis and hyperhidrosis.² It must be differentiated from other conditions, such as congenital hemangioma, infantile hemangioma, vascular malformations, pyogenic granuloma and, in adults, kaposiform hemangioendothelioma and Kaposi sarcoma. To differentiate between other tumors or to assess the area involved, imaging as ultrasound or magnetic resonance can be used.³ Histopathology of TA shows many lobules of tufts spread across the dermis with a “cannonball” appearance, crescent-shaped spaces around the vascular tufts and similar spaces in the tumor stroma.^{2,4,5} Immunohistochemistry can be strongly positive for *Ulex uropaeus* I lectin and EN4, besides CD31 and CD34, and rarely positive for smooth muscle actin and negative staining for GLUT1.^{3,4} The main treatment option for tufted angioma is surgical excision. Other therapeutic modalities have been reported, such as cryotherapy, laser, topical or systemic corticosteroids and chemotherapy. Some authors believe the lesion should only be monitored due to the possibility of spontaneous regression of these cases.^{3,5} □

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MAILING ADDRESS:

Camila Mendes da Silva
R. Codajás, 24
Cachoeirinha
69065-130 Manaus, AM
E-mail: mendes_camila@hotmail.com

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Effectiveness and safety of infliximab for 11 years in a patient with erythrodermic psoriasis and psoriatic arthritis*

Lívia Arroyo Trídico¹
João Roberto Antonio^{1,2}
Carlos Eduardo Mathias¹
Eurides Maria de Oliveira Pozetti^{1,2}

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Dear Editor,

Psoriasis is a chronic skin condition that affects 0.5 to 5% of the population. It is characterized by erythematous, scaly plaques, typically affecting the extensor surfaces of the knees and elbows, scalp, intergluteal cleft and sacrum.¹ Around 40% of psoriasis patients have psoriatic arthritis, which leads to physical limitations, decreased quality of life, and increase in patient mortality.^{2,3}

Erythrodermic psoriasis is a rare form of psoriasis, characterized by generalized erythema with variable scaling. It is associated with severe morbidity and even mortality, since it can cause hyper or hypothermia leading to decreased cardiac output and affecting liver and renal functions. Erythrodermic psoriasis results from worsening of a previous case of psoriasis or it can develop as the initial presentation of psoriasis. Its treatment is still a challenge and is not yet standardized due to lack of scientific evidence regarding therapeutic recommendations. Traditional systemic therapies include methotrexate, cyclosporine and oral retinoid.^{4,5,6}

Tumor necrosis factor alpha (TNF- α) is a key mediator in the pathogenesis of psoriasis, because it is involved in keratinocyte proliferation, endothelial cell regulation and T lymphocyte recruitment.³ Based on the important role of TNF- α in the pathogenesis of psoriasis, the first biologic drugs emerged, which changed radically the treatment for patients with moderate to severe psoriasis.² Infliximab was the first biologic drug used for treatment of psoriasis, approved by the Food and Drug Administration (FDA) in 2006.⁷

Erythrodermic psoriasis treatment with traditional systemic medications (methotrexate, cyclosporine and oral retinoid), although effective, is frequently associated with therapeutic failure or intolerance, with the need of alternative strategies. Recently, TNF

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¹ Dermatology Service of the Hospital de Base da Faculdade de Medicina de São José do Rio Preto (FAMERP) - São José do Rio Preto (SP), Brazil.

² Discipline of Dermatology of the Faculdade de Medicina de São José do Rio Preto (FAMERP) - São José do Rio Preto (SP), Brazil.