

CASES FOR DIAGNOSIS

Reticular Telangiectatic Plaque

A Alfaro-Rubio, C Serra-Guillén, and R Botella-Estrada

Servicio de Dermatología, Instituto Valenciano de Oncología, Valencia, Spain

Patient History

A 21-year-old woman consulted for lesions on the back of the neck that were slightly painful to touch and had lasted 1 year. She reported that the lesions were in an area where she had undergone radiotherapy for Kasabach-Merritt syndrome that had developed over a congenital vascular tumor diagnosed as a cavernous hemangioma. Biopsy had been taken of the lesions on 2 occasions, with a histological diagnosis of capillary angioma. The patient was treated with pulsed dye laser, wavelength 595 nm, 2-ms pulse duration, and 8 J/cm² energy, with no improvement in the lesions or pain. One month after laser treatment, the lesions had spread to the side of the neck and the supraclavicular region.

Physical Examination

The physical examination revealed a vascular lesion composed of pale red telangiectatic macules located on the back and side of the neck (Figure 1A) and the right supraclavicular area (Figure 1B). Some of these macules

were confluent and formed a discontinuous reticular plaque. The plaque borders were imprecise and had areas that disappeared with pressure.

Additional Examinations

Doppler ultrasound and magnetic resonance imaging showed findings consistent with a low-flow vascular malformation, probably of venous origin. The histopathological study showed an intact epidermis, with clusters of cells scattered through the superficial and deep dermis in cannonball fashion; most were accompanied by dilated vessels (Figure 2). At higher magnifications, the clusters were seen to be associated with the vessel walls, displacing the lumen and producing half-moon images. These tufts were formed by densely organized endothelial cells with no atypias, which left groove-like vascular spaces between them, some of which were packed with red blood cells (Figure 3).

What is your diagnosis?

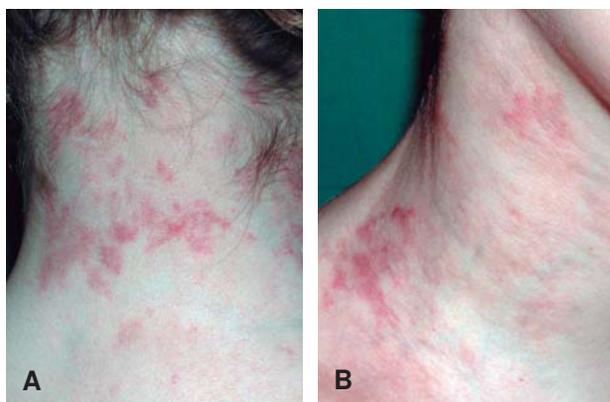


Figure 1.

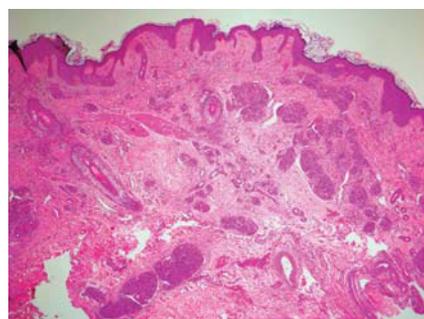


Figure 2.
Hematoxylin-eosin, ×10.

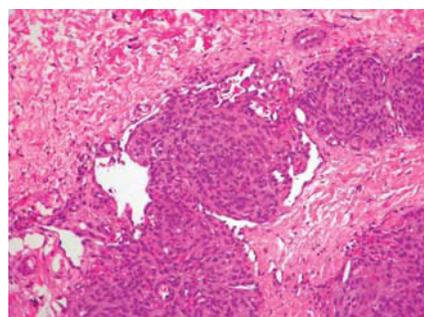


Figure 3.
Hematoxylin-eosin, ×20.

Correspondence:
Alberto Alfaro Rubio
Servicio de Dermatología. Instituto Valenciano de Oncología
Profesor Beltrán Báguena, 8, 46009 Valencia, Spain
aalfaro@aedv.es

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Diagnosis

Tufted angioma

Comment

Tufted angioma is a rare benign vascular tumor previously known as progressive capillary hemangioma or angioblastoma.¹ Its etiology is unknown. Although onset is usually before age 5 years, it has developed during adulthood in some cases and more rarely, has appeared in congenital form.² Two cases of familial inheritance have been described.³

Tufted angioma manifests clinically as coalescent purplish-red papules that form plaques usually located on the neck, upper portion of the thorax, or shoulders. The natural course of the lesion is usually slow, progressive growth at first, with stabilization afterwards, although several cases of spontaneous regression have also been reported.⁴

A rare complication of tufted angioma is Kasabach-Merritt phenomenon, which consists of a large vascular tumor associated with thrombocytopenic coagulopathy.⁵ The treatment for Kasabach-Merritt phenomenon associated with tufted angioma may leave residual lesions that can have 3 different clinical patterns: type I, red macules with or without papules (this pattern may vary over time and may be associated with pain); type II, raised telangiectatic streaks; and type III, palpable irregular mass, demonstrable by imaging techniques. On occasions, the residual tumor is the one that redevelops slowly, with varying signs and symptoms, including pain.⁶

There is little information available on the effective treatment of tufted angioma. Pulsed dye laser causes selective vascular damage in the skin with minimal epidermal damage and, therefore, is the treatment of choice for vascular lesions.⁷ Tufted angioma has a varied response to this kind of treatment, from lightening of the color of the lesion⁸ to lack of response.⁹ Pulsed dye laser is usually effective in

decreasing the pain caused by the tumor^{8,9}; however, in some cases, it is only able to reduce the coloration of the lesion.⁸

Conflicts of Interest

The authors declare no conflicts of interest.

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