

a circumscribed morphology. Postoperative alopecia is due to hypoxia caused by pressure on the reclined head during a long general anesthetic. Alopecia areata has a more complex and multifactorial etiology and pathogenesis. In the majority of cases, the diagnosis of transitory rectangular alopecia after endovascular embolization is based on the history of radiation to the area and the artifactual morphology of the lesions (typically rectangular). However, the absence of inflammation and the presence of dystrophic hair shafts can make it difficult to distinguish the condition from alopecia areata.¹ In these cases, histopathology of a biopsy from the affected area is useful to differentiate the 2 entities. From a histopathological point of view, transitory rectangular alopecia after endovascular embolization should theoretically show changes compatible with anagen effluvium, associated with a minimal inflammatory infiltrate, although the findings will depend to a large extent on the moment at which the biopsy is performed. In our case, the biopsy showed a minimal inflammatory infiltrate and all the follicles were in catagen after the abrupt cessation of their cycle. We did not observe a honeycomb-like morphology of the infiltrate around the follicular bulb, typical of alopecia areata, or signs of radiodermatitis in the interfollicular epidermis or in the follicular epithelium. The absence of areas of scarring confirmed the transitory nature of the alopecia, as was observed clinically.

With the progressive increase in the number and complexity of endovascular embolization techniques, treating ever more complex lesions and requiring longer fluoroscopy times,⁵⁻⁷ it is likely that consultations for transitory rectangular alopecia after endovascular embolization will increase in the near future. It is therefore important that dermatologists are aware of the clinical characteristics and histopathologic features of this condition, so that they can reassure patients and inform them that the clinical course will be favorable.

References

1. Tosti A, Piraccini BM, Alagna G. Temporary hair loss simulating alopecia areata after endovascular surgery of cerebral arteriovenous malformations: A report of 3 cases. *Arch Dermatol.* 1999;135:1555-6.
2. Freysz M, Mertz L, Lipsker D. Temporary localized alopecia following neuroradiological procedures: 18 cases. *Ann Dermatol Venereol.* 2014;141:15-22.
3. Balter S, Hopewell JW, Miller DL, Wagner LK, Zelefsky MJ. Fluoroscopically guided interventional procedures: A review of radiation effects on patients' skin and hair. *Radiology.* 2010;254:326-41.
4. Garnacho GM, Amorrinch MV, Salido R, Espejo J. Alopecia rectangular transitoria tras embolización de aneurisma: a propósito de un caso. *Actas Dermosifiliogr.* 2009;100:907-22.
5. Wen CS, Lin SM, Chen Y, Chen JC, Wang YH, Tseng SH. Radiation-induced temporary alopecia after embolization of cerebral arteriovenous malformations. *Clin Neurol Neurosurg.* 2003;105:215-7.
6. Podlipnik S, Giavedoni P, San-Román L, Ferrando J. Square alopecia: A new type of transient alopecia of the scalp following fluoroscopically endovascular embolization. *Int J Trichology.* 2013;5:201-3.
7. César A, Baudrier T, Mota A, Azevedo F. Geometric alopecia after embolization of a ruptured aneurysm. *Actas Dermosifiliogr.* 2015;106:148-50.

C. Bernárdez, I. Alcaraz, A.M. Molina-Ruiz,* L. Requena

Servicio de Dermatología, Hospital Universitario Fundación Jiménez Díaz, Universidad Autónoma, Madrid, Spain

* Corresponding author.

E-mail address: amolinar@fjd.es (A.M. Molina-Ruiz).

Giant Vascular Eccrine Spiradenoma[☆]



Espiradenoma ecрино vascular gigante

To the Editor,

Giant vascular eccrine spiradenoma is a rare variant of eccrine spiradenoma characterized by its size (> 2 cm diameter) and high vascularity, a feature that often leads to it being confused with tumors of vascular origin. We present a case of this rare tumor.

A 54-year-old woman presented with a tumor that had been present for years, measuring 2.3 × 2 × 2.6 cm, located on the medial aspect of the left arm (Fig. 1A). The tumor was a reddish, dome-shaped lesion, ulcerated on one side,



Figure 1 A, Cutaneous component with a dome-like pedunculated vascular appearance and ulceration at the superior pole. B, MRI showing an exophytic multilobulated mass with a blood supply from and drainage to the basilic vein (arrow).

[☆] Please cite this article as: Jorquera Barquero E, Lara Bohórquez C, de Alba Rioja I. Espiradenoma ecрино vascular gigante. *Actas Dermosifiliogr.* 2015;106:850-852.

Table 1 Clinical Characteristics of Giant Vascular Eccrine Spiradenomas.

Reference	Age/Sex	Size, cm	Anatomic Location	Clinical Diagnosis
Cotton et al ¹	74/male	5	Abdomen	Angiosarcoma
	84/female	2	Occipital scalp	Sebaceous cyst
Hey et al ²	63/female	3.5 × 1.5	Thigh	Venous thrombosis
Senol et al ³	60/male	3-4	Chest	Angioma or thrombosis
Ko et al ⁴	56/female	2	Back	Angiolipoma or neuroma
Yamakoshi et al ⁵	76/male	5 × 3.4 × 2.6	Shoulder	Not stated
Kim et al ⁶	49/female	2.5 × 2.5	Arm	Vascular malformation
Tremezaygues et al ⁷	81/male	1.5 × 2.6 × 2.9	Forearm	Not stated
Hatano et al ⁸	52/female	3.5 × 2.2	Arm	Pilomatricoma
Krishnan et al ¹⁰	31/male	2.5	Chest	Not stated
Nokamura et al ⁹	79/male	7 × 7 × 6	Chest	Chronic expansive hematoma
Moragon Gordon et al	60/female	5 × 6	Breast	Not stated
Our case	54/female	2.3 × 2 × 2.6	Arm	Hemangioma

and hard to palpation. The patient attended because the mass had increased in size and at some point there had been a small amount of bleeding. Magnetic resonance imaging (MRI) was performed to investigate the clinical suspicion of a tumor of vascular origin. The different projections (sagittal plane on T2; transverse plane on T1, T2, FFE T2, and STIR) showed a mass located in the subcutaneous cellular tissue, which was presumed to be of vascular origin given that it was described as being dependent on and draining into the basilic vein (Fig. 1B). The lesion was mottled and multilobulated. There was no intramuscular component and the surrounding tissues were unchanged. On the basis of these findings, we proceeded to surgical excision and histopathological study. The tumor, situated in the mid-dermis and deep dermis, was composed of several well-demarcated lobules separated by fine bands of fibrous tissue with abundant dilated and congested vascular structures (Fig. 2). The lobules were arranged in aggregates of cells, some with a basaloid appearance and others with a larger, clearer cytoplasm, arranged in cords, trabeculae, and solid nests with duct-like structures. On immunohistochemical study, most of the cells expressed pan cytokeratins, and the luminal cells expressed carcinoembryonic antigen (CEA).

Spiradenomas are rare benign tumors derived from the inferior portion of eccrine sweat ducts, and they usually develop on the head, neck, trunk, and, less often, the limbs. Their main clinical characteristic is that they are generally painful. Typically, spiradenomas present in patients aged between 15 and 35 years of age and they are usually solitary, although different clinical presentations have been described.

In 1986, Cotton et al¹ described two cases of eccrine spiradenoma that they classified as giant vascular type due to their large size and high vascularity. Since then, to the best of our knowledge, 12 further cases have been described (Table 1). The first description referred to a classic spiradenoma, measuring over the established 2 cm diameter, with the most salient feature being its high vascularity. Many authors have described this high vascularity as the result of a stromal involution process due to ageing of the lesion.^{7,9} This concept is defended by some authors,⁹ but in our case the information supplied by MRI supported the existence of a tributary vessel. This vessel made it easy to mistake the mass

for a vascular tumor, and it would also have promoted tumor growth. The most notable feature in this case was the finding of an afferent vessel to the tumor with drainage into the basilic vein, leading the radiologist to suspect a slow flow vascular malformation. Similarly, the marked vascularization of these tumors means that most eccrine spiradenomas are clinically diagnosed as benign or malignant neoplasms

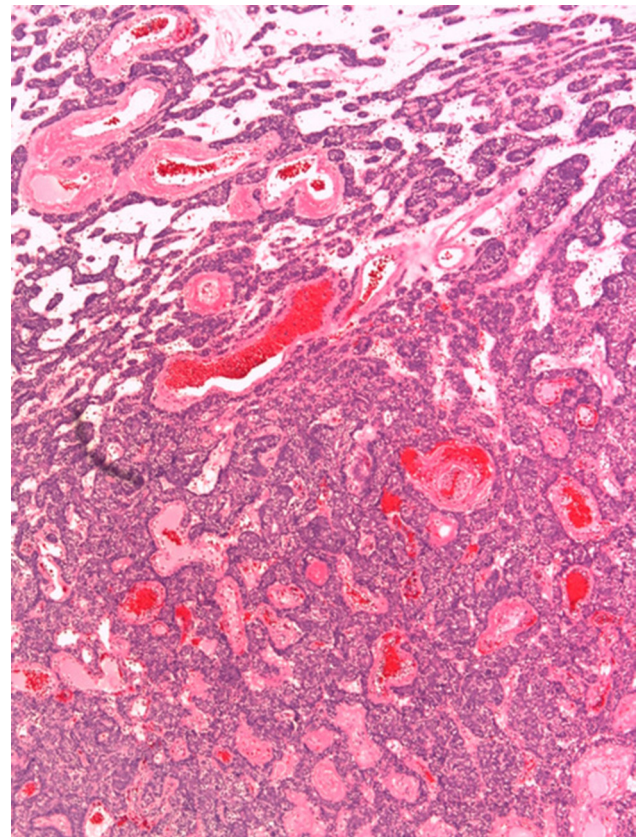


Figure 2 Organoid pattern of cell nests, some of basaloid appearance with hyperchromatic nuclei and others with large clear cytoplasm, separated by fibrous tracts containing abundant dilated and congestive vascular structures (hematoxylin-eosin × 10).

of vascular origin. The most important differential diagnoses are angiosarcoma, which expresses endothelial markers such as CD31 and CD34, and chronic expansive hematoma, which presents as an encapsulated tumor filled with blood and neovasculature.

What was interesting about this case, apart from the rarity of the condition, was the ease with which a giant vascular eccrine spiradenoma could be mistaken for a lesion of vascular origin.

References

1. Cotton DWK, Slater DN, Rooney N, Goepel JR, Mills PM. Giant vascular eccrine spiradenomas: A report of two cases with histology, immunohistology and electron microscopy. *Histopathology*. 1986;10:1093–9.
2. Hey A, Grouls V, Röckelein G. Vascular eccrine giant spiradenoma-A case report with histology and immunohistology of a rare variant of benign sweat gland tumors. *Z Hautkr*. 1988;3:444–7.
3. Senol M, Ozcan A, Sasmaz S, Ozen S, Ciralik H. Giant vascular eccrine spiradenoma. *Int J Dermatol*. 1998;37:221–3.
4. Ko JY, Lee CW, Moon SH, Song KW, Park CK. Giant eccrine spiradenoma: Report of a case with immunohistochemical study. *J Korean Med Sci*. 2006;21:172–6.
5. Yamakoshi T, Makino T, Watanabe H, Furuichi M, Matsui K, Shimizu T. A case of giant vascular eccrine spiradenoma with unusual clinical features. *Clin Exp Dermatol*. 2009;34:e250–1.
6. Kim MH, Cho E, Lee JD, Cho SH. Giant vascular eccrine spiradenoma. *Ann Dermatol*. 2011;19:197–200.
7. Tremezaygues LL, Pfohler C, Vogt T, Muller CS. Differential considerations of skin tumors with florid vascularisation: Report of a solitary giant vascular eccrine spiradenoma. *BMJ*. 2011, <http://dx.doi.org/10.1136/bcr.05.2011.4187>.
8. Hatano A, Kobayashi T, Fukuzumi S, Shirakashi Y, Nishimura C, Gomi H, et al. Giant vascular eccrine spiradenoma mimicking calcifying epithelioma. *J Dermatol*. 2013;40:853–4.
9. Nakamura Y, Teramoto Y, Yamada K, Yamamoto A. Giant vascular eccrine spiradenoma mimicking chronic expanding hematoma. *J Dtsch Dermatol Ges*. 2014;10:908–10.
10. Krishnan K, Thomas A. Giant vascular eccrine spiradenoma with unusual clinical presentation: A case report. *Open J Pathol*. 2014;4:21–4.

E. Jorquera Barquero,^{a,*} C. Lara Bohórquez,^b
I. de Alba Rioja^a

^a Servicio Dermatología M-Q y Venereología, Complejo hospitalario de Huelva, Huelva, Spain

^b Servicio de Anatomía Patológica, Complejo hospitalario de Huelva, Huelva, Spain

* Corresponding author.

E-mail address: jorroc@aedv.es (E. Jorquera Barquero).

Refractory Hailey-Hailey Disease That Responded Well to Photodynamic Therapy[☆]



Enfermedad de Hailey-Hailey recalcitrante con buena respuesta a terapia fotodinámica

To the Editor:

Hailey-Hailey disease or familial benign pemphigus is a rare hereditary skin disease. This chronic and recurrent condition can cause severe discomfort and be difficult to control with conventional treatments. Photodynamic therapy (PDT) could offer an alternative in cases in which other treatment options have failed.

The patient was a 56-year-old male who for 30 years had experienced several outbreaks a year of stinging, itching, and burning lesions located on the neck and armpits. The lesions interfered with his daily activities and had a negative impact on his quality of life. Physical examination revealed bright, well-defined, highly exudative, eroded, and erythematous plaques with some surface crusts located in both armpits and on the lateral aspects of the neck (Fig. 1). Histological study of a biopsied sample from one of the lesions demonstrated an epidermis with erosions

and intraepidermal blisters, marked acantholysis, and the typical appearance of a dilapidated brick wall. The patient had been diagnosed with Hailey-Hailey disease and had received multiple treatments with astringents such as copper and zinc sulfate (1:1000), topical and systemic steroids, topical vitamin D derivatives, topical tacrolimus, and acitretin. Response to treatment had always been partial with a disease-free interval between flares lasting only a few weeks. These shortcomings represented an important limitation for the patient. He agreed to undergo a single session of PDT to see what the response would be and to assess the tolerability of the treatment. First, both underarms were cleaned with physiological saline solution and methyl aminolevulinate (MAL) cream was applied under occlusion for 3 hours. The underarms were then irradiated with a red light (Aktilite at 37 J/cm²) for 7.5 minutes. During the period of exposure, the patient experienced a slight sensation of pain and burning, which was well tolerated. The response on follow-up at 2 weeks was excellent: the erosions had healed, the exudate and erythema had disappeared, and the patient reported an improvement in his quality of life (Fig. 2). Six months later, the patient is still free of lesions and has not undergone any other treatment.

Hailey-Hailey disease or familial benign pemphigus is an autosomal dominant genodermatosis caused by a mutation in the *ATP2C1* gene. It is characterized by the appearance of

[☆] Please cite this article as: Lobato-Berezo A, Imbernón-Moya A, Aguilar-Martínez A. Enfermedad de Hailey-Hailey recalcitrante con buena respuesta a terapia fotodinámica. *Actas Dermosifiliogr*. 2015;106:852–854.