# UC Davis
## Dermatology Online Journal

### Title
Superficial angiomyxoma of the nipple: a case report of an infrequent cutaneous tumour

### Permalink
https://escholarship.org/uc/item/9mk7m2tp

### Journal
Dermatology Online Journal, 22(10)

### Authors
Victoria Martínez, Ana Mercedes
Sánchez Carazo, José Luis
Alegre de Miquel, Victor

### Publication Date
2016

### DOI
10.5070/D32210032899

### Copyright Information
Copyright 2016 by the author(s).This work is made available under the terms of a Creative Commons Attribution-NonCommercial-NoDerivatives License, available at https://creativecommons.org/licenses/by-nc-nd/4.0/

Peer reviewed

eScholarship.org Powered by the California Digital Library
University of California

**Volume 22 Number 10**
**October 2016**

**Case Presentation**

# Superficial angiomyxoma of the nipple: a case report of an infrequent cutaneous tumour

**Ana Mercedes Victoria Martínez, José Luis Sánchez Carazo, Victor Alegre de Miquel**

**Dermatology Online Journal 22 (10): 8**

**General University Hospital of Valencia, Valencia, Spain**

**Correspondence:**

Ana Mercedes Victoria Martínez
Email: anamercevictoria@gmail.com

## Abstract

Superficial angiomyxoma is a distinctive cutaneous soft tissue neoplasm that is clinically variable, infrequent, and benign. However, this tumor has a high propensity for local recurrence. There is a known association of angiomyxomas with Carney complex. We report a case of superficial angiomyxoma in a 28-year-old woman who presented with an erythematous, multilobulated nodule on the nipple. This tumor has a predilection for the trunk, head and neck, extremities, and genitalia. The present case, hence, is unusual, because the lesion developed on the nipple. The appearance of a solitary cutaneous angiomyxoma warrants an examination to rule out the presence of Carney complex. After clinical examination, echocardiogram and endocrine analyses, Carney complex was ruled out in our patient.

**Keywords: Carney complex; myxoid tumors, superficial angiomyxoma**

## Introduction

Superficial angiomyxomas (SAM) are rare, clinically variable, benign skin lesions that can be locally aggressive and recur if they are not completely removed [1]. It was first described in 1988 as an entity that is histologically and clinically indistinguishable from cutaneous myxomas arising in Carney complex [2]. Although SAM is a distinct entity [3], the appearance of a solitary cutaneous angiomyxoma warrants an examination to rule out the presence of Carney complex.

### Case synopsis

A 28-year-old previously healthy woman presented with a 1-year history of a solitary nodule on the left nipple. The lesion was a 12x8-mm in size, tender, erythematous, multilobulated nodule (Figure 1).

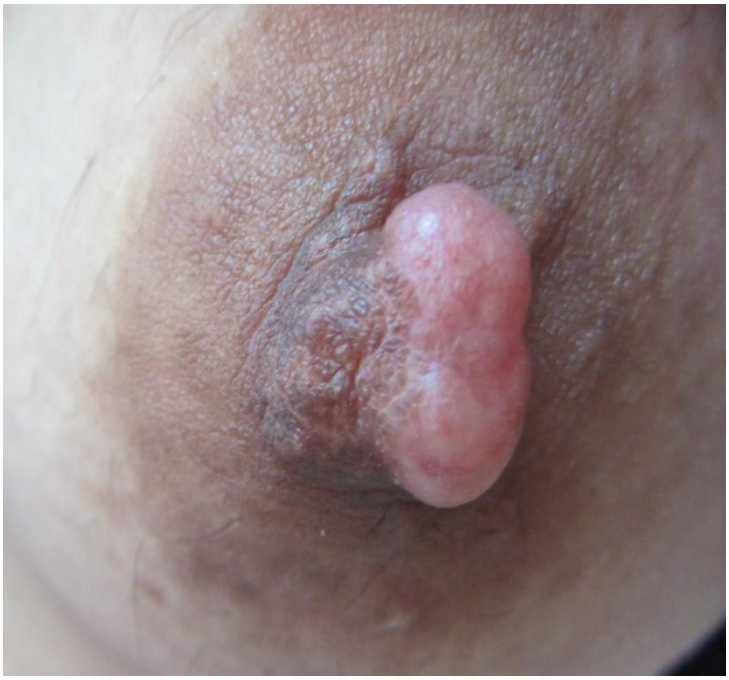

**Figure 1.** Erythematous multilobulated nodule on the left nipple.

She also reported some mild pain in the nipple during menstruation but denied other associated symptoms. The patient had no other similar lesions and had no ephelides, lentigines, or other pigmentation of the skin or mucous membranes. The lesion was surgically excised under local anesthesia. Histopathologic examination revealed a multilobulated nodule in the dermis (Figure 2), with proliferation of stellate and round to spindled cells with relatively uniform nuclei and scant cytoplasm embedded in myxoid stroma. It showed a scattered distribution of small thin-walled blood vessels and inflammatory cells.

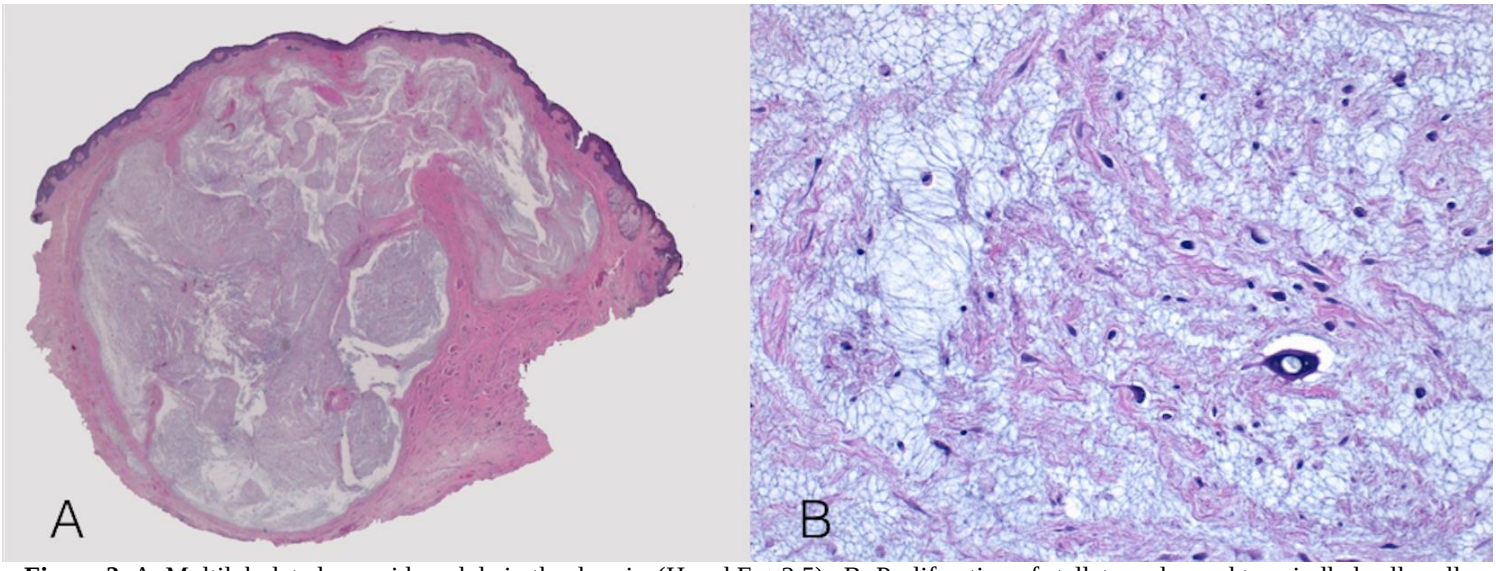


**Figure 2.** A, Multilobulated myxoid nodule in the dermis, (H and E, ×2.5) , B, Proliferation of stellate and round to spindled cells cells embedded in myxoid stroma (H&E, ×40).

Immunohistological stains were positive for vimentin and negative for S100 and actin. Although the patient had no evidence of Carney complex, an echocardiogram and laboratory tests, including a complete blood cell count and comprehensive metabolic panel, were done and were all normal. At one year follow up after the excision there was no evidence of local recurrence.

## Discussion

Superficial angiomyxomata are typically solitary skin nodules or polypoid lesions less than 5 cm in size. This tumor has a predilection for the trunk, head and neck, extremities, and genitalia. The present case is unusual, because the lesion developed on

the nipple. When SAM is multiple or is present on the external ear [4], it is almost pathognomonic for Carney complex. SAM is more frequent in men and occurs most commonly in the third to fourth decade.

Histologically, SAM demonstrates a poorly demarcated tumor centered in the dermis with common extension into subcutaneous fat. This is characterized by a myxoid stroma with a well-defined proliferation of spindle, stellate, and oval fibroblasts and prominent vascularization consisting of small, thin-walled, blood vessels. An inflammatory cell component, including interstitial neutrophils, is usually seen. In a third of cases an epithelial component (epithelial strands or epidermoid cysts) is identified [5]. It is positive for vimentin and negative for S100, with occasional positivity for CD34. These histological features help differentiate SAM from other myxoid tumors, such as myxoid neurothekeoma, which demonstrates positive staining for S100, absence of neutrophils in the infiltrate, and focal cutaneous mucinosis. Generally, a well-defined dermal mucin accumulation with thin collagen fibers and without the presence of a reticulin network or elastic fibers is demonstrated. There is a tendency of local recurrence [6].

There is a known association of angiomyxomas with Carney complex, a rare autosomal dominant multiple neoplasia syndrome, which includes the presence of cardiac and cutaneous myxomas, lentigines, epithelioid blue nevi, psammomatous melanocytic schwannomas, osteochondromyxomas, endocrine hyperactivity, and pituitary tumors [7]. The tumor suppressor gene PRKAR1A is found to be mutated in almost 60% of Carney Complex cases [8].

Complete excision and follow-up observation are recommended because superficial angiomyxomas present a high propensity for local recurrence which occurs in 30% to 40% of patients and is associated with inadequate resection [1]. Some authors defend the use of Mohs surgery using permanent section control for treatment of SAM in cosmetically sensitive areas [10].

# References


1. Allen PW, Dymock RB, MacCormac LB. Superficial angiomyxomas with and without epithelial components. Report of 30 tumors in 28 patients. Am J Surg Pathol. 1988;12:519–30. [PMID: 3389450]
2. Carney JA, Headington JT, Su WP. Cutaneous myxomas. A major component of the complex of myxomas, spotty pigmentation, and endocrine overactivity. Arch Dermatol. 1986;122:790–8. [PMID: 3729510]
3. Calonje E, Guerin D, McCormick D, Fletcher CD. Superficial angiomyxoma: Clinicopathologic analysis of a series of distinctive but poorly recognized cutaneous tumors with tendency for recurrence. Am J Surg Pathol. 1999;23:910–7. [PMID: 10435560]
4. Ferreiro JA, Carney JA. Myxomas of the external ear and their significance. Am J Surg Pathol. 1994;18(3):274-280. [PMID: 8116795]
5. Shanthi V, Krishan BAR, Rao NM, Sekhar Reddy R, Poorna Chandra Sekhar BP, Harish CV. Superficial angiomyxoma with epithelial component—a rare cutaneous myxoid tumor. J Biosci Tech. 2011;2(3):318-323.
6. Allen PW. Myxoma is not a single entity: a review of the concept of myxoma. Ann. Diagn. Pathol. 2000; 4: 99–123. [PMID: 10760324]
7. Broker SS, Kamath SG,Kashyap N, Sagar SC, Rao L, Warrier R et al. Carney complex:case report and review. J Cardiothorac Surg. 2011;6:25. [PMID: 21375769]
8. Rothenbuhler A, Stratakis CA. Clinical and molecular genetics of Carney complex. Best Pract Res Clin Endocrinol Metab. 2010;24(3):389-399.[PMID: 20833331]
9. Rocco F, Cozzi G, Spinelli MG, Rocco BM, Albo G, Finkelberg E, et al. Massive recurring angiomyxoma of the scrotum in a obese man. Rare Tumors 2011; 3: 98–9. [PMID: 22066038]
10. Kahn SL, Juhl ME, Sidiropoulos M, Guitart J, Antonijevic S, Krunic AL. Angiomyxoma of the nasal dorsum treated by Mohs surgery. Australas J Dermatol. 2014 Oct 21. [PMID: 25333656]