World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Eccrine Spiradenoma

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World Journal of Advanced Research and Reviews, 2024, 22(01), 586–590

Publication history: Received on 28 February 2024; revised on 07 April 2024; accepted on 09 April 2024

Article DOI: https://doi.org/10.30574/wjarr.2024.22.1.1113

Abstract

First described in 1956, spiradenomas are benign tumors that arise from the bulge of the hair follicle rather than the sweat gland.

Epidemiologically, spiradenomas are rare, malignant ones are even rarer, with only around 120 documented cases worldwide.

No racial association or sexual predilection for spiradenomas has been reported.

Most spiradenomas occur between the ages of 15 and 35. As for malignant spiradenomas, they tend to develop after the age of 50, with a median age of 60 years.

Clinically, they are generally skin-colored nodules approximately 1 cm in diameter with a soft consistency, located on the scalp, neck, upper part of the trunk, breasts and rarely the armpits and vulva.

While malignant spiradenomas tend to have slightly different characteristics from benign spiradenomas which can metastasize with an estimated rate of 19%.

We report the case of a 45-year-old primiparous patient who had been postmenopausal for 5 years and had a maternal aunt who died of breast cancer who consulted for the discovery on self-palpation of a subcutaneous nodule near the left axillary area.

The breast examination is unremarkable on D cup breasts with palpation of a subcutaneous nodule measuring 2.5 x 1.5 cm near the left axillary area.

Echo mammography finds tissue and cystic lesions of the left breast classified ACR3 and a left axillary subcutaneous lesion measuring 24x13mm that is difficult to characterize.

After biopsy of this lesion, the histological and immunohistochemical appearance returned in favor of a cutaneous adnexal tumor compatible with an ECCRINE SPIRADENOMA

The extension assessment made by Thoracal abdomino pelvic CT had revealed non-specific pulmonary micronodules to be monitored

The patient was sent to multidisciplinary team discussion with the decision to perform a left lumpectomy of the axillary lesion.

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The management of malignant spiradenomas is multidisciplinary with a surgeon, an oncologist and possibly a radiation oncologist.

Keywords: Breast; Biopsy; Histology; Spiradenomas; Imaging

1. Introduction

Spiradenomas are benign tumors originally thought to arise from the sweat glands of the dermal layer as they were first described in 1956 by Kersting and Helwig, naming them eccrine spiradenomas (1).

In current practice, they are considered to originate from the bulge of the hair follicle rather than the sweat gland.

2. Case report

We report the case of a 45-year-old primiparous patient who had been menopausal for 5 years and had a maternal aunt who died of breast cancer who consulted following the discovery on self-palpation of a subcutaneous nodule near the left axillary area.

The breast examination is unremarkable on D cup breasts with perception of a subcutaneous nodule measuring 2.5 x 1.5cm near the left axillary area (image 1).

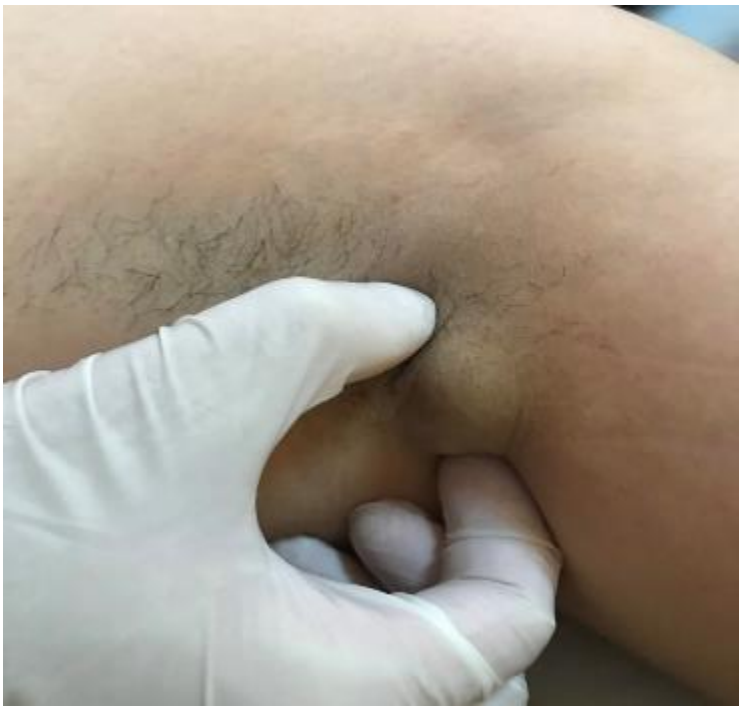

Figure 1 Physical examination of our patient

The echo mammogram found tissue lesions in both breasts classified as ACR3, the result of the pathological examination was a fibroadenoma; and a left axillary subcutaneous lesion measuring 24x13mm that is difficult to characterize. The latter's biopsy came back in favor of adenoid cystic carcinoma.

The breast MRI did not add any more information.

The extension assessment carried out by Thoracal abdominal pelvic CT showed non-specific pulmonary micronodules to be monitored.

The patient was then assigned to multidisciplinary team discussion with the decision to perform a left lumpectomy of the axillary lesion.

Definitive anatomopathology study concluded in eccrine spiradenoma (image2).

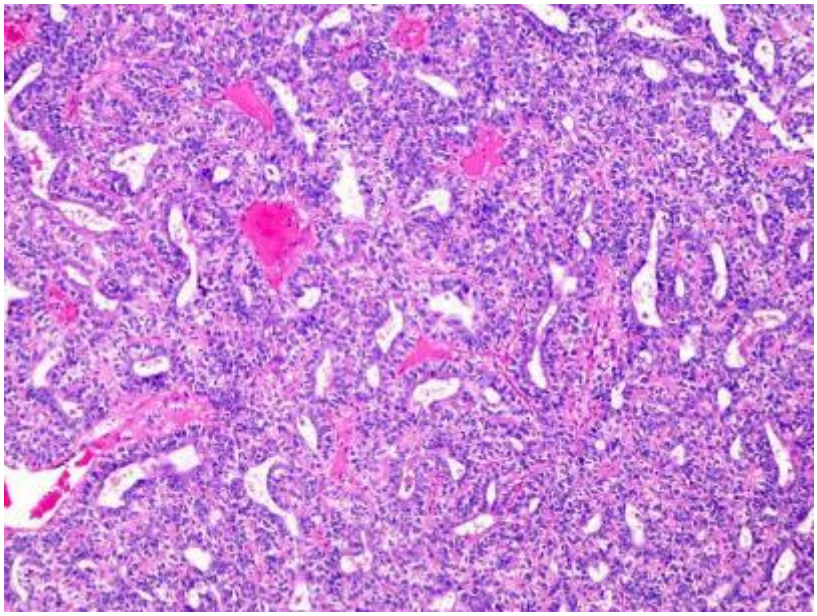

Figure 2 Histology of eccrine spiradenoma

3. Discussion

3.1. Epidemiology

Spiradenomas are rare , with the exact incidence of benign solitary spiradenomas unknown. Malignant spiradenomas are even rarer, with only about 120 documented cases worldwide.

Even rarer are cases of variable morphologies of multiple spiradenomas, with fewer than 40 cases documented in the literature (2).

Most spiradenomas occur in people aged 15 to 35. They can occur at any age, with reports of a spiradenoma occurring at 4 weeks of age.

Malignant spiradenomas tend to develop after age 50, with a median age of 60 years. There have been rare reports of malignant transformation of spiradenomas in children.

There is no racial link or sexual predilection for spiradenomas or malignant spiradenomas (3).

3.2. Physiopathology

In pathophysiology, it is a defective tumor suppressor gene that leads to the development of spiradenomas (4).

In Brooke-Spiegler syndrome, of which spiradenomas are a manifestation, the defective gene is CYLD on chromosome 9.

The gene product of CYLD downregulates the NF-κB signaling pathway, and when this protein is nonfunctional, increased activity of the NF-κB pathway leads to uncontrolled cell proliferation, resulting in the development of adnexal tumors (5). .

3.3. Physical examination

Clinically, spiradenomas are usually skin-colored, gray, pink, purple, red, or blue nodules approximately 1 cm in diameter. Lesions tend to occur on the scalp, neck, upper trunk, breasts, and rarely the armpits and vulva (6). Spiradenomas tend to be soft and are sometimes tender to palpation.

Malignant spiradenomas tend to have slightly different characteristics than benign spiradenomas based on size and appearance. They tend to be larger than benign spiradenomas, with a median diameter at presentation of 3 cm (range, 0.8-25 cm). Malignant spiradenomas also tend to ulcerate. They can also metastasize (7).

A review of spiradenocarcinomas found a metastasis rate of 19%, with the most common sites being the lungs, bones and lymph nodes. Less common locations of metastases were the liver, kidney and breast (8). Thus, it is important to evaluate for potential metastatic disease during a physical examination. The mortality rate in case of metastasis is 10%.

3.4. Imaging

Several imaging methods can be used such as ultrasound, MRI or CT but especially in the case of a malignant spiradenoma to locate metastases.

Spiradenoma should be considered in the differential diagnosis of a hypervascularized and hypoechoic subcutaneous tumor that is well circumscribed on ultrasound (9).

It is usually located in the subcutaneous fat layer and has a well-defined margin. The appearance is usually hypoechoic, lobulated, and may resemble a path with a heterogeneous echo texture. Other reported features include internal hypoechoic foci with peripheral blood flow (10).

Malignant spiradenomas appear on MRI as multiple dispersive foci with clear circumferences in the skin and subcutaneously. They demonstrate low signal intensity on T1- type images and high signal intensity on inversion recovery images (11).

3.5. Anatomo-pathology

- Skin biopsy is necessary to establish the diagnosis of spiradenoma.
- Surgical excision is the treatment of choice (9).
- Fine-needle cytology of a breast spiradenoma may be performed to aid in the diagnosis.

3.6. Treatment and care

The basis of treatment for benign spiradenomas is surgical excision. The lesions do not tend to recur after surgery. Mohs micrographic surgery was used on the anterior neck to surgically remove the persistent infiltrating spiradenoma.

Multiple spiradenomas, such as those found in Brooke-Spiegler syndrome, were treated with a high-energy continuous-wave carbon dioxide laser after reduction with bipolar scissors in one patient (12).

The management of malignant spiradenomas is multidisciplinary with a surgeon, an oncologist and possibly a radiation oncologist.

Complete excision of the lesions must be carried out to prevent recurrence with a 1 cm safety margin (13).

Surgery is generally curative; therefore, additional care is not always necessary. Patients with malignant spiradenomas should be followed by oncologists and radiation oncologists (14).

Radiotherapy and chemotherapy can be used after tumor resection of malignant spiradenomas

There is no medical treatment for spiradenomas. However, treatment of malignant spiradenoma may include tamoxifen-based chemotherapy (12, 14).

4. Conclusion

Eccrine spiradenoma is a rare tumor, developed on the epithelium of eccrine sweat glands.

Malignan form is possible and the features of the examination are important to think about it.

Surgery is recommended and is generally the key of treatment.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

[1] Sellheyer K. Spiradenoma and cylindroma originate from the hair follicle bulge and not from the eccrine sweat gland: an immunohistochemical study with CD200 and other stem cell markers. *J Cutan Pathol*. 2014 Oct 29.

[2] Dabska M. Malignant transformation of eccrine spiradenoma. *Pol Med J*. 1972. 11(2):388-96.

[3] Bumgardner AC, Hsu S, Nunez-Gussman JK, Schwartz MR. Trichoepitheliomas and eccrine spiradenomas with spiradenoma/cylindroma overlap. *Int J Dermatol*. 2005 May. 44(5):415-7.

[4] Clarke J, Ioffreda M, Helm KF. Multiple familial trichoepitheliomas: a folliculosebaceous-apocrine genodermatosis. *Am J Dermatopathol*. 2002 Oct. 24(5):402-5.

[5] Sun SC. CYLD: a tumor suppressor deubiquitinase regulating NF-kappaB activation and diverse biological processes. *Cell Death Differ*. 2010 Jan. 17 (1):25-34.

[6] Grossmann P, Vanecek T, Steiner P, et al. Novel and Recurrent Germline and Somatic Mutations in a Cohort of 67 Patients From 48 Families With Brooke-Spiegler Syndrome Including the Phenotypic Variant of Multiple Familial Trichoepitheliomas and Correlation With the Histopathologic Findings in 379 Biopsy Specimens. *Am J Dermatopathol*. 2013 Feb. 35(1):34-44.

[7] Im M, Kim DH, Park JS, Chung H, Lee Y, Kim CD, et al. Alteration of the β-catenin pathway in spiradenoma. *J Cutan Pathol*. 2011 Aug. 38 (8):657-62.

[8] Rashid M, van der Horst M, Mentzel T, et al. ALPK1 hotspot mutation as a driver of human spiradenoma and spiradenocarcinoma. *Nat Commun*. 2019 May 17. 10 (1):2213

[9] Englander L, Emer JJ, McClain D, Amin B, Turner RB. A rare case of multiple segmental eccrine spiradenomas. *J Clin Aesthet Dermatol*. 2011 Apr. 4(4):38-44.

[10] Kazakov DV, Schaller J, Vanecek T, Kacerovska D, Michal M. Brooke-Spiegler syndrome: report of a case with a novel mutation in the CYLD gene and different types of somatic mutations in benign and malignant tumors. *J Cutan Pathol*. 2010 Aug. 37(8):886-90.

[11] Bowen S, Gill M, Lee DA, Fisher G, Geronemus RG, Vazquez ME, et al. Mutations in the CYLD gene in Brooke-Spiegler syndrome, familial cylindromatosis, and multiple familial trichoepithelioma: lack of genotype-phenotype correlation. *J Invest Dermatol*. 2005 May. 124 (5):919-20.

[12] Staiger RD, Helmchen B, Papet C, Mattiello D, Zingg U. Spiradenocarcinoma: A Comprehensive Data Review. *Am J Dermatopathol*. 2017 Oct. 39 (10):715-725.

[13] Tiradogonzalez M, Beierle E, Hammers Y, Andea A, Mroczek E. Neonatal Spiradenoma. *Pediatr Dermatol*. 2012 Jul 2.

[14] Miedema JR, Burgon E, Burkhart C, Stitzenberg K, Hipps J, Zedek D. Metastatic spiradenocarcinoma occurring in an 8-year-old boy. *Pediatr Dermatol*. 2015 Jan-Feb. 32 (1):122-7.