Benign eccrine spiradenoma imitating a nerve sheath tumor: illustrative case

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BACKGROUND Benign eccrine spiradenoma is a rare tumor arising from the sweat glands and is a pathology that is almost never encountered in routine neurosurgical practice. Although this is a rare pathology, it is one that should be included in the differential diagnosis for a patient presenting with a painful, subcutaneous mass, because it can guide further treatment considerations.

OBSERVATIONS The authors present a case of benign eccrine spiradenoma that mimicked a nerve sheath tumor in clinical presentation, imaging characteristics, and gross appearance.

LESSONS Complete local excision of these lesions is the gold standard treatment, because they are painful, and there are reports of local recurrence and malignant degeneration with incomplete resection. For this reason, neurosurgeons should be sure to include this in the differential diagnosis of a patient with a painful, subcutaneous mass, because it may help to guide management decisions.

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KEYWORDS eccrine spiradenoma; nerve sheath tumor; peripheral nerve

Benign eccrine spiradenoma is a tumor that arises from the sweat glands in the dermis or subcutaneous tissue, most commonly in young adults. It is a pathology that is not commonly encountered in routine neurosurgical practice, but it can mimic pathologies that are, such as nerve sheath tumors (NSTs). We present a case of a benign eccrine spiradenoma in the arm of a woman.

Illustrative Case

Clinical Presentation

A 45-year-old female with no significant medical history presented for evaluation of an enlarging, painful, subcutaneous mass on the dorsal aspect of the right upper arm. There was concern that this mass could be a melanoma, because the patient had a positive family history, causing her to present to surgical oncology for evaluation. After evaluation by the surgical oncologist, the mass was thought to be most consistent with an NST, and she was referred to neurosurgery. The patient reported that the mass had been present for years but had recently increased in size. It was tender to touch. In addition, she endorsed intermittent numbness and tingling radiating down the arm from the mass and into the fingers on the right hand. Her neurological examination in clinic was unremarkable, with full strength and intact sensation throughout the right upper limb. Her examination was notable for a visible and palpable subcutaneous mass in the dorsal aspect of the right upper arm, with tenderness to palpation over the mass, as well as a positive Tinel sign.

Imaging Characteristics

Ultrasound of the mass revealed a predominantly solid, hypoechoic mass with smooth borders. There were small cystic areas within the mass (Fig. 1). Magnetic resonance imaging (MRI) was then obtained for further characterization, which redemonstrated a 1.0 × 1.0 × 1.5-cm, bilobed, subcutaneous mass with homogeneous enhancement on T1-weighted postcontrast sequences. The mass was hypointense on T1-weighted MRI scans and hyperintense on T2-weighted MRI scans. The mass was in close proximity to the expected course of the superior lateral brachial cutaneous nerve, a small branch arising from the axillary nerve. The appearance on MRI was thought to be most consistent with benign NST versus melanoma (Fig. 2).
Cal marginal involvement but no evidence of malignancy. The stromal changes noted, as well as a scattered ductal structure (Fig. 3).

Postoperative Course
Consultation with a surgical oncologist and radiation oncologist at our institution confirmed the rare nature of this tumor. A recommendation was made for no further treatment and surveillance only. The patient was seen in follow-up 2 weeks postoperatively and reported complete resolution of her symptoms.

Discussion
Observations
Eccrine spiradenoma is a rare neoplasm that was first described by Kersting and Helwig in 1956. It arises from eccrine sweat glands in the dermis and superficial subcutaneous tissue. Usually, there is no connection to the overlying epidermis. It can occur at any age but most commonly affects young adults, and it has no apparent sex predilection. A large majority of patients present with pain or tenderness associated with the lesion, as was seen in 91% of the patients described by Kersting and Helwig and in our patient as well. Clinically, the tumors appear as firm, well-circumscribed masses, most commonly involving the face, neck, and trunk, predominantly on the ventral surface, although there are case reports of lesions involving the extremities. As of 2017, there were only 25 reported cases of eccrine spiradenoma arising from the upper or lower limbs.

Ultrasound evaluation of the mass generally reveals a smooth, lobulated mass in the superficial subcutaneous tissue. The mass appears hypoechoic and often has increased vascularity. The classic MRI appearance is a T1-hypointense, T2-hyperintense mass, with homogeneous enhancement after contrast administration.

Gross examination of the tumors generally reveals a firm, round or ovoid mass, sometimes with small cystic components. The color can vary from gray to pink, brown, or yellow or some combination of these. Characteristic histological findings on H&E staining are two distinct cell types: a basaloid cell population with hyperchromatic nuclei and scant cytoplasm and a larger cell type with abundant, pale cytoplasm. The cells are arranged into sheets, cords, or trabeculae, with the paler cells more centrally located. Some tumors are markedly vascular with cystic dilation of their vascular channels, likely accounting for their increased vascularity seen on ultrasound.

Although most cases of spiradenoma are benign, they can recur locally with incomplete excision. In addition, there are just over 100 cases reported of malignant transformation. Malignant spiradenomas nearly always arise from a long-standing benign lesion, although they can rarely arise as de novo malignancies. Malignant spiradenomas tend to be diagnosed in older patients. They can present with rapid growth, erythema, ulceration, and bleeding and may resemble melanoma. Pathology shows anaplastic basaloid cells with areas of highly pleomorphic cells and numerous mitotic figures. Because of their potential for local recurrence or malignant degeneration, complete surgical excision is the preferred treatment method for the benign variant of the tumor, with routine follow-up to screen for local recurrence. Our patient was offered a second surgery with wider surgical margins, but she preferred surveillance.

Lessons
This case represents a unique presentation of a pathology not commonly encountered by neurosurgeons. The clinical, MRI, and gross appearance of the mass were consistent with a benign NST.
but the final pathology serves as a reminder that other, rarer pathologies must be considered in the differential diagnosis of similar lesions. Differential considerations include benign NST (schwannoma or neurofibroma), traumatic neuromas, and malignant NST. See Table 1 for characteristic imaging and histological findings for these types of lesions.

In conclusion, we report this interesting case of a very rare pathology, eccrine spiradenoma mimicking an NST, as a reminder to include this unique pathology in the differential diagnosis when evaluating a patient with a painful subcutaneous mass.

References
1. Kersting DW, Helwig EB. Eccrine spiradenoma. AMJ Dermatol 1956;73(3):199–227.
2. Mambo NC. Eccrine spiradenoma: clinical and pathologic study of 49 tumors. J Cutan Pathol. 1983;10(5):312–320.
3. Son JH, Choi YW, Cho YS, et al. A Case of Eccrine Spiradenoma: A Rarely Seen Soft Tissue Tumor on the Extensor Surface of Arm. Ann Dermatol. 2017;29(4):519–522.
4. Kwon KE, Kim SJ, Choi HJ, et al. Sonographic appearance of an eccrine spiradenoma: A case report. J Clin Ultrasound. 2018;46(7):494–496.
5. Han YD, Huan Y, Deng JL, Zhang YG, Zhang CH. MRI appearance of multiple eccrine spiradenomas. Br J Radiol. 2007;80(949):e27–e29.
6. Andreoli MT, Itani KMF. Malignant eccrine spiradenoma: a meta-analysis of reported cases. Am J Surg. 2011;201(5):685–699.
7. Murphey MD, Smith WS, Smith SE, Kransdorf MJ, Temple HT. Imaging of musculoskeletal neurogenic tumors: radiologic-pathologic correlation. Radiographics. 1999;19(5):1283–1280.
8. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: diagnostic overview and update on selected diagnostic problems. Acta Neuropathol. 2012;123(3):295–319.
9. Golan JD, Jacques L. Nonneoplastic peripheral nerve tumors. Neurosurg Clin N Am. 2004;15(2):223–230.

Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Hanna. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Bowman. Critically revising the article: all authors. Reviewed submitted version of manuscript: Hanna, Bowman. Approved the final version of the manuscript on behalf of all authors: Hanna. Administrative/technical/material support: Hanna. Study supervision: Hanna.

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TABLE 1. Differential diagnosis for a painful subcutaneous mass in the upper extremity

| Tumor Type       | Gross Appearance                                                                 | MRI Appearance                                                                 | Pathological Findings                                                                 |
|------------------|----------------------------------------------------------------------------------|--------------------------------------------------------------------------------|--------------------------------------------------------------------------------------|
| Schwannoma       | Well-circumscribed, firm, yellow-tan mass                                         | Isointense to hyperintense on T1, hyperintense on T2; homogeneous enhancement³   | Biphasic appearance (Antoni A & B) w/ “school of fish” arrangement of spindle cells; Verocay bodies²⁸ |
| Neurofibroma      | Well-circumscribed, white-gray mass                                               | Similar to schwannoma, “target sign” on T2²                                    | Wavy, elongated cells w/ “shredded carrot” type collagen²⁸                                |
| Malignant PNST    | Fleshy, variegated mass w/ necrosis & hemorrhage                                   | Similar to benign PNST, but w/ infiltration of surrounding tissue, can see evidence of necrosis/hemorrhage³ | Hypercellular spindle cell population w/ palisading necrosis & mitotic figures³          |
| Traumatic neuroma | Bulbous or fusiform mass arising at the end of or within an injured nerve          | Isointense on T1, isointense to hyperintense on T2; may contrast enhance³        | Tangled proliferation of axons & Schwann cells; nonneoplastic⁷                             |
| Eccrine spiradenoma| Round, well-circumscribed, firm mass, w/ occasional cystic component¹             | Hypointense on T1, hyperintense on T2; homogeneous contrast enhancement⁵         | Basaloid cells mixed w/ larger, paler cells & lymphocytic infiltrate¹²                   |

PNST = peripheral nerve sheath tumor.