Painful Eccrine Spiradenoma Containing Nerve Fibers: A Case Report

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Abstract
An eccrine spiradenoma is a rare benign tumor most often seen in the head, neck and upper trunk of young adults. Although spontaneous pain or tenderness is a typical symptom of eccrine spiradenomas, the underlying mechanism has not been fully elucidated. Here, we report the case of a 47-year-old woman who had a spiradenoma in the subcutaneous tissue of her posterior neck accompanied by agonizing pain which was triggered by pressure. Multiple nodular lesions were excised and the typical histopathological findings of spiradenoma were seen. The histopathological architecture of a disorganized nerve fiber encasing the tumor nodules appeared to correlate with the unique clinical symptom of pain.

Introduction
A spiradenoma is an uncommon benign tumor arising from the eccrine sweat apparatus and is one of nine painful skin tumors, the others being leiomyoma, neuromatoma, dermatofibroma, angiolipoma, neurilemmoma, endometrioma, glomus tumor and granular cell tumor [1]. The generation of pain is thought to be related to small unmyelinated axons permeating the hyalinized stromal mantle [2]. Some investigators believe that the expansion of cysts in the tumor also gives rise to pain [3]. Although the pain of an eccrine spiradenoma is ‘the most frequent and striking symptom’, which is seen in 91% of reported patients [4], the mechanism of pain generation has not been elucidated fully. We experienced a patient with spiradenoma who had severe pain triggered by palpation or pressure, which was treated with surgical excision. The unique clinical characteristics of the pain and the surgical and histopathological findings are depicted. The peculiar microscopic architecture of the tumor nodules and nerve fibers in the capsule might explain the pain mechanism of a spiradenoma.

Case Report

History and Examinations
A 47-year-old woman visited the outpatient clinic with a posterior neck mass causing fierce pain that was evoked by pressure. The texture of the skin surface was not different from other areas, except for a slight elevation. The mobile soft mass was barely palpable over the spinal column near the midline. The agonizing pain was triggered only when the mass was palpated with a certain amount of pressure. No pain was generated by a soft touch or mild pressure. Once pressure triggered the pain, it persisted for approximately 1–2 min in the absence of pressure. The patient described the pain as sharp, similar to an electrical shock, and radiating to the posterior head and both shoulders; it was so severe that she felt like fainting.

The patient had had the lesion for about 20 years. The initial symptom was an intermittent spontaneous pricking sense that occurred several times per day, but was not related to touch or pressure. The pricking sensation changed into pain that was triggered by pressure about 12 years earlier. These pains had caused her to visit a local clinic and undergo magnetic resonance imaging (MRI) 11 years earlier. Although the MRI showed subtle lesions, the doctor did not relate the pain to the lesion.

The patient always asked people to avoid touching her posterior neck. Despite these precautions, she experienced agonizing pain inevitably or unexpectedly, such as when examined by a doctor at another hospital, when scrubbed by a professional...
scrubber in a public bathhouse, or when someone swatted at a bug on her neck. Whenever a doctor examined her, she experienced fierce pain, which made her evade definitive treatment. Because no visible lesion was detected on her skin surface, we initially suspected some abnormality or tumorous condition in the deep structure and requested cervical spine MRI. On MRI, multiple well-margined ovoid masses were seen in the subcutaneous fat on her posterior neck (fig. 1). She decided to eliminate the source of pain and requested general anesthesia for fear of pain.

**Surgical Findings and Procedures**

Under general anesthesia, a 4 × 1 cm spindle-shaped vertical skin incision was made over the mass. Eight ovoid or irregularly shaped masses measuring 3–10 mm were removed from the subcutaneous fat (fig. 2). Each yellow-pink mass was encapsulated with an isolated thin fibrous membrane and easily separated from the surrounding fat. Fine vasculature was seen on some of the surfaces. A wedge resection of the subcutaneous tissue around the mass was performed to remove any remaining tiny lesions. Her postoperative course was uneventful and the pain disappeared.

**Pathology**

Microscopic examination of the histological sections showed sharply delineated solitary strongly basophilic nodules, resembling lymph nodes. The tumor nodules were surrounded by a delicate fibrous capsule, some of which contained blood vessels and prominent thickened nerve fibers (fig. 3). The S-100 protein and neuron-specific enolase revealed positivity in the thickened nerve fibers (fig. 4). The tumor was made up of two cell types, the most frequent being large cells with a large pale nucleus and a nucleolus. The nuclear membrane was thin and distinct, and the chromatin was finely and evenly distributed. The cytoplasm was pale to slightly basophilic. The second cell type was smaller and had a hyperchromatic nucleus and scanty cytoplasm. Neither type of cell showed atypia. This organoid pattern of the epithelial cells was interrupted by dilated vascular channels. Lymphocytes were scattered throughout the tumor. The cells were arranged in intertwining bands interspersed with small lumens (fig. 5). Immunohistochemically, the tumor cells expressed pancytokeratin and CK7 (fig. 6).

**Discussion**

An eccrine spiradenoma is a rare tumor of the eccrine sweat glands and was first described by Kersting and Helwig [4] in 1956. In their study of 134 eccrine spiradenomas, more than 97% of the tumors were solitary blue-red dermal or subcutaneous nodules, ranging from 0.5 to 3 cm in diameter. Eccrine spiradenoma occurs as either solitary lesion or as multiple lesions. In some case, the multiple lesions present in a linear or zosteriform distribution [5–7]. In the current case, multiple tumor nodules were found in subcutaneous tissue. Although it is unclear whether the current case is a single tumor with many lobules or multiple spiradenomas, we favor single tumor with multiple lobules because several nodules were com-

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**Fig. 1.** MRI: T2-weighted sagittal (a) and axial (b) images showed multiple ovoid masses with discrete margins in the subcutaneous fat tissue of the posterior neck.
Fig. 2. a–c Grossly, the surgical specimen contained 8 ovoid or irregularly shaped masses measuring 3–10 mm in subcutaneous fat. The thinly encapsulated yellow-pink masses were easily separated from the surrounding fat. Fine vasculature was found on some of their surfaces.

Fig. 3. a The tumor nodules are surrounded by a delicate fibrous capsule, some of which contain blood vessels and prominent thickened nerve fibers. HE. ×40. b A high-power view of a capsule area containing nerve fibers. HE. ×200.
The tumor cells are arranged in intertwining bands and numerous lumens. HE. ×100.

The tumor cells are positive for pancytokeratin. Immunohistochemistry. ×200.

Fig. 4. Immunohistochemically, the nerve fibers in the capsule express S-100 protein (a) and neuron-specific enolase (b). Immunohistochemistry. ×100.

The most striking clinical feature of the lesions was the presence of pain or tenderness in 91% of the patients, which usually occurs in a paroxysm [4]. In contrast, Mambo [8] reported that pain or tenderness was present much less frequently (only 23%) in his clinical and histopathological review of 49 cases of eccrine spiradenoma.

The differential diagnosis should include leiomyoma, neuroma, dermatofibroma, angiolipoma, neurilemmoma, endometrioma, glomus tumor and granular cell tumor, described as 'LEND AN EGG' by Naversen [1]. The exact mechanism of pain in most of the painful skin tumors is not clear. In the case of a leiomyoma, which is also among the known painful skin tumors, the pain mechanism was proven to be mediated by the contraction of smooth muscle.
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References


