

Spiradenoma: A Case Report with Evidence Reinforcing the Thickened Nerve Fibers in the Tumor Capsule

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Authors' contributions

This work was carried out in collaboration between all authors. Author SPK wrote the draft of the manuscript and provided the case. Author MK managed the literature searches. Authors NK and AA designed the pathological figures, managed literature searches. Author CA contributed to the writing and correction of the draft. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aim: Spiradenoma is a rare tumor. Spontaneous pain and tenderness on palpation is a frequent symptom of the disease. Although the cause of pain in spiradenoma is not clarified yet, thickened nerve fibers in the capsule of tumor have been suggested to be related to the pain symptoms.

Case: We here report a woman with painful nodules on her abdomen for two months. The histopathological examination confirmed the diagnosis of spiradenoma. Immunohistochemically the tumor nodule and capsule were both S-100 positive, whereas the neuron specific enolase stained only the capsule of the tumor.

Discussion: Although further studies are required to explain the exact mechanism of pain in these

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tumors, by reporting this case, we provide additional evidence for the existence of thickened nerve fibers in the capsule of spiradenoma.

Keywords: Spiradenoma; painful skin tumor; nerve fibers; S-100; neuron specific enolase.

1. INTRODUCTION

Spiradenoma is a rare tumor thought to originate from eccrine sweat glands, though some lesions may demonstrate apocrine differentiation. Spontaneous pain and tenderness on palpation is a frequent symptom of the disease. Although the cause of pain in spiradenoma is not clarified yet, thickened nerve fibers in the capsule of tumor have been suggested to be related to the pain symptoms [1].

2. CASE

The patient was a 39-year-old woman who had a 2-month history of painful-to-touch nodular lesions on her abdomen. On physical examination, multiple linearly arranged tender red-blue nodules 0.2-0.5cm in diameter were observed on the left side of her abdomen [Fig. 1].

No other cutaneous abnormalities were present. A biopsy was taken from the biggest nodule. The histopathological examination showed sharply demarcated basophilic tumor lobules located in the dermis with lack of connection to the epidermis [Fig. 2a].

There were two kinds of tumor cells: small cells with dark nuclei at the periphery and larger pale cells near the lumen. The basaloid cells were arranged in a trabecular pattern with eosinophilic

fibrous strands and large cells show luminal differentiation containing eosinophilic material [Fig. 2b].



Fig. 1. Firm, tender, red-blue small nodules linearly arranged

Immunohistochemically S-100 protein positivity was observed both in the parenchyma and capsule of the tumor [Fig. 3a]. Nerve fibers in the capsule express neuron specific enolase [Fig. 3b].

No signs of malignancy were present. The diagnosis of spiradenoma was made. The patient refused total excision. She was seen 2 years later with lesions slightly enlarged [Fig. 4].

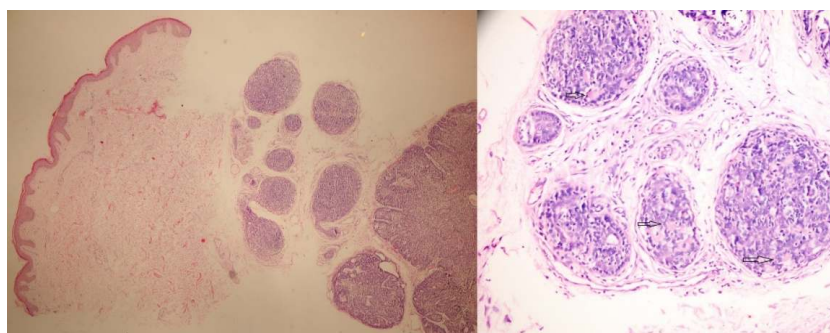


Fig. 2a. The appearance of 'blue balls in the dermis' The nodules are well margined with a fibrous capsule. The interlacing cords with evidence of ductal differentiation (HEx40), b. The basaloid cells are arranged in a trabecular pattern with eosinophilic fibrous strands and large cells show luminal differentiation containing eosinophilic material (arrows) (HEx100)

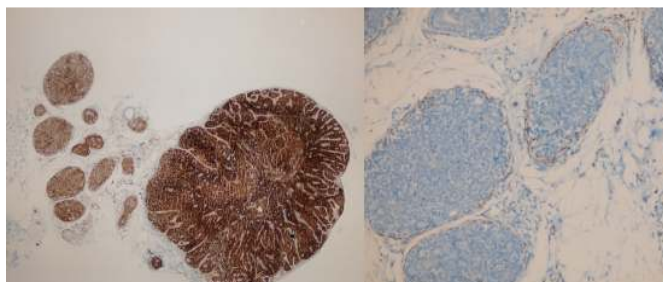


Fig. 3a. S-100 protein positivity are seen in the parenchyma and capsule of the tumor (S-100x100), b. The nerve fibers in the capsule express neuron specific enolase (NSEx100)



Fig. 4. Lesions slightly enlarged within two years

3. DISCUSSION

Spiradenoma was first described in 1956 by Kersting and Herwig as an uncommon tumor of the eccrine sweat glands [2]. Most of the cases are solitary painful nodules seen on head, neck and upper trunk of young adults with a male/female ratio of 1:3. Multiple lesions may arise in association with Brooke-Spiegler syndrome which is an autosomal dominant condition of multiple cylindromas, trichoepitheliomas and cylindromas [1]. In our patient no other cutaneous abnormalities were present. Non-syndromic spiradenoma might have multifocal or focal distribution and have rarely been reported to have zosteriform, linear, nevoid or blaschkoid pattern [3]. The histopathology of spiradenoma is characterized by multinodular, sharply-circumscribed neoplasms of basaloid cells within the dermis and subcutis. Because of their blue color, they are described as 'blue balls' in the dermis. Within the nodules aggregates of two cell types are seen as small, basaloid cells with hyperchromatic nuclei and large cells with pale nuclei often located closer to the center. The duct-like structures, eosinophilic hyaline material pseudoglandular rosettes might be seen as an evidence of ductal differentiation [4]. Malignant

transformation of spiradenoma may occur in case of rapidly enlarging nodule with a histopathology including atypical cells with frequent mitoses. The histopathological examination of our case revealed no signs of malignancy and the luminal differentiation containing eosinophilic material was representative of benign nature. Indeed, the lesions only slightly enlarged within two years. Treatment option of spiradenoma is surgical excision if possible. The diagnosis depends on typical clinical feature and the histopathological examinations which are quite characteristic.

Spontaneous pain and tenderness on palpation is a frequent symptom of spiradenoma. The differential diagnosis of painful skin tumors should include leiomyoma, eccrine spiradenoma, neuroma, dermatofibroma, angioliipoma, neurilemmoma, endometrioma, glomus tumor and granular cell tumor described as 'LEND AN EGG' mnemonic [1-5]. However, several other skin lesions or neoplasms also can be added to the list of these painful tumors. Several mechanisms have been described to explain the cause of pain in these tender cutaneous tumors. As one might expect, tumors can elicit pain by invading nerves or as a mass effect by compressing the neighboring nerve fibers. Again, enlarging tumors with subsequent necrosis may elicit pain by driving inflammation. Even the chemical mediators secreted from tumors are suggested to stimulate sensory nerve endings and cause pain symptoms. However, the mechanisms of spontaneous pain caused by tumors are mostly not clear [6]. Considering leiomyoma, the pain is proposed to result from the contraction of smooth muscle fibers or local compression of nerve endings [7]. The pain mechanism of a neuroma which occurs by the formation of regenerative axon sprouts after trauma is thought to be in neuropathic origin. The injured afferent nerve fiber itself has been shown to become sensitive to mechanical, thermal, and

chemical stimuli and cause pain symptoms [8]. Similarly, peripheral nerve sheath tumors which includes granular cell tumor and dermal schwannoma [neurilemmoma] are proposed to trigger pain by increased axonal transmission due to ectopic mechanosensitivity or successive self stimulation of nerve fibers which undergo aberrant axonal sprouting [9]. Although the cause of pain in spiradenoma is yet not fully understood, thickened nerve fibers in the capsule of tumor nodules have been suggested to be related to the pain symptoms [1]. Previously, the compression of nerve fibers found in the tumor capsule was suggested as the cause of pain related to angioleiomyoma [10]. Park et al. [1] reported a case of spiradenoma with prominent thickened nerve fibers in the tumor capsule. The tumor capsule was S-100 protein and neuron-specific enolase positive immunohistochemically but the tumor nodule was S-100 negative. In line with their finding, we observed S-100 and neuron-specific enolase positivity in the tumor capsule. However, the tumor nodule was also S-100 positive in our case. The staining of tumor nodule with S-100 was not surprising in that it points the eccrine differentiation of the tumor as stated in previous cases [5].

4. CONCLUSION

Although we are far away from confirming an etiological relation between the pain symptoms and the nerve fiber involvement in these tumors, our case strenghtens the finding of neural elements in the capsule of eccrine spiradenoma. Further studies are necessary to define the role of nerve involvement in the pain symptoms of these tumors.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this paper and accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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