CASE REPORT
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VULVAR NEURILEMMOMA AT THE LABIUM MINUS

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Neurilemmoma is a benign slow-growing nerve sheath neoplasm that is rarely found in the female genital system. To date, thirteen cases of vulvar neurilemmoma have been reported in the world literature. We report here on the first Korean case of vulvar neurilemmoma. A 43-year-old woman presented with a 0.6×0.6 cm sized oval nodule at the left labium minus. Complete excision was done. Microscopically, the mass was a well circumscribed lesion that was alternatively composed of cellular Antoni A and paucicellular Antoni B areas; The Antoni A areas were composed of spindle cells with twisted, buckled nuclei and occasional intranuclear vacuoles, and the spindle cells were arranged in short bundles or fascicles. Immunohistochemically, the spindle cells were positive for S100 protein and negative for desmin. Although it has a very low incidence, neurilemmoma should be considered in the differential diagnosis of a cutaneous or subcutaneous mass in the vulva.

Keywords: Neurilemmoma; Vulva; S100 proteins

Neurilemmoma is also named neurinoma, schwannoma and Schwann cell tumor. It is generally a solitary, nodular, benign tumor that arises from the neural sheath Schwann cells of the peripheral, cranial or autonomic nerves. The most common locations are the head, neck, upper and lower extremities, posterior mediastinum and retroperitoneum [1]. It rarely affects the female genitalia. To the best of our knowledge, epithelial tumors are the most common tumor in the vulvar region, and mesenchymal tumors comprise less than 10% of them. Neurilemmoma involving vulva is the least common. To date, only thirteen cases of vulvar neurilemmoma have been reported in the literature [2-14]. Here, we report on an additional case of vulvar neurilemmoma arising in the labium minus in a patient without neurofibromatosis.

Case Report

A 43-year-old Korean woman (gravida 3, para 2) presented with a non-tender palpable small nodule at her left labium minus. The patient reported that she noticed the small nodule in her left vulvar region 2 weeks previously. The size measured 0.6×0.5×0.4 cm. Her previous menstrual history was regular. On general examination, she was found to be of average build and there were no clinical signs indicative of neurofibromatosis 1 and 2. On gynecologic examination, there was no abnormality except the small nodule. A simple excision of the nodule was performed under local anesthesia. After 6 months, she has had no evidence of recurrence or symptoms.

1. Pathologic findings

The excised nodule was situated in the lower part of the dermis and it was a well-circumscribed encapsulated lobular nodule for the architecture (Fig. 1A). Nuclear palisadings that formed Verocay bodies were found in the Antoni A area, and in the Antoni B areas, the cells were loosely arranged within a slightly myxoid stroma (Fig. 1B). The tumor cells were spindle shaped with nuclear buckling. The nucleoli were not prominent, and mitosis or necrosis was not seen. Perivascular hyalinization was also noted. Immunohisto-

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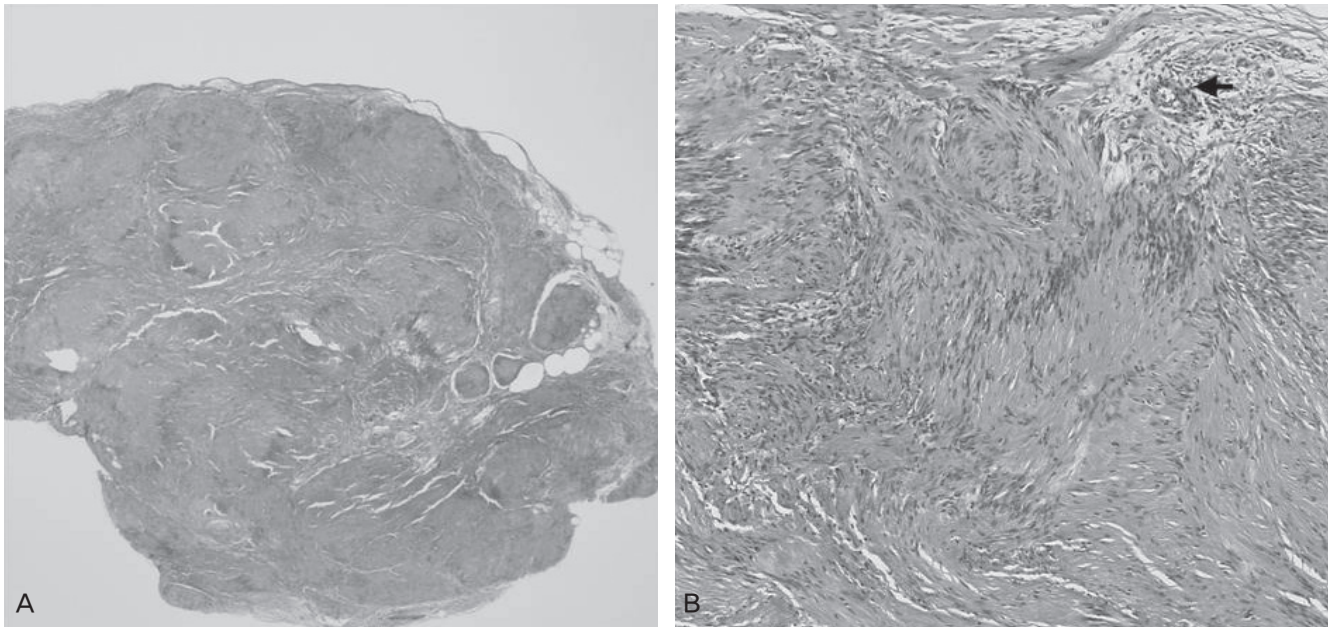

Fig. 1. (A) Low power view reveals a well circumscribed subcutaneous mass (H&E, ×40). (B) In a densely cellular Antoni A area, nuclear palisadings with Verocay bodies are seen. Note the hyalinized blood vessels are found in a paucicellular Antoni B area (arrow, H&E, ×200).

chemically, the spindle cells were strongly positive for S100 protein (polyclonal; 1:1200 dilution, Zymed, San Francisco, CA, USA), and they were negative for pancytokeratin (AE1/AE3; Dako, Glostrup, Denmark, prediluted), desmin (D33; Dako, 1:100 dilution) and smooth muscle actin (1A4; Dako, 1:100 dilution). Neurilemmoma arising in the labium minus was diagnosed.

Discussion

Neurilemmoma is an encapsulated, well circumscribed tumor that is classically characterized by cellular Antoni A and paucicellular Antoni B areas. The conventional type is the most common and the plexiform type with multinodular growth and cellular variants or ancient variants, i.e., degenerative changes can also be found [1]. Neurilemmoma is a tumor that arises from the Schwann cells of the nerve sheath, and it occurs throughout the whole body. Neurilemmoma can arise from neurofibromatosis or it can occur sporadically. Of all the neurogenic tumors, about half are found in the head and neck region. The origin of neurilemmoma is most commonly associated with a nerve trunk, and the clinical symptoms depend on the nerve of origin. Vulvar neurilemmoma is a slow-growing well encapsulated benign tumor. After reviewing the previous literature about vulvar neurilemmoma, the vulvar neurilemmoma presents with nonspecific symptoms that are indistinguishable from those of other cutaneous lesions; the lesions had presented for many years with a slow increase in size [2-14]. Due to the site of occurrence, sitting and walking can be affected, but most of the vulvar neurilemmoma are discovered by chance. Histologically, the conventional type of neurilemmoma is the most common type in the vulvar area. It was interesting in that plexiform (multinodular) neurilemmoma is one of the least common histologic variants of neurilemmomas, but five cases of plexiform variants and a case of ancient variant have been found among the reported cases of vulvar neurilemmoma [2,4,8,11-13]. The age at presentation of a vulvar neurilemmoma has varied from 5 to 84 years. The size of vulvar neurilemmoma ranged from 0.6 cm up to 15 cm (mean, 4.3 cm). Most of the vulvar neurilemmoma occurred in the labium except for four cases of clitoral neurilemmoma [3,9,12,13]. None of the reported patients with vulvar neurilemmoma were associated with neurofibromatosis, and even for the patients with plexiform neurilemmoma. These findings are summarized in Table 1.

The nature of vulvar neurilemmoma has not yet been properly characterized in the previously reported cases including the present one. In general, neurilemmoma has a true capsule composed of epineurium, which makes the nerve be displaced to the periphery by the mass when large nerves are the sites of origin. This finding is helpful, but it is not found in neurilemmoma of the small nerves. Growth of neurilemmoma within the epineurium creates

Table 1. Review of the previously reported cases of the vulvar neurilemmoma including the present one

No	Authors	Year	Age	NF	Pathologic subtype	Clinical outcome
1	Huang et al. [3]	1983	84	–	Conventional	NR
2	Woodruff et al. [4]	1983	26	–	Plexiform	2 recurrences
3	Yamashita et al. [5]	1996	29	–	Conventional	NR
4	Hanafy et al. [6]	1997	59	–	UK	NR
5	Quesada et al. [7]	1998	68	–	Conventional	NR
6	Santos et al. [8]	2001	5	–	Plexiform	NR
7	Llaneza et al. [9]	2002	64	–	Conventional	NR
8	Fujimoto et al. [10]	2004	22	UK	Conventional	UK
9	Agaram et al. [11]	2005	26	–	Plexiform	NR
10	Chuang et al. [12]	2007	41	–	Plexiform	NR
11	Yegane et al. [13]	2008	6	–	Plexiform	NR
12	Das et al. [14]	2008	48	–	Conventional	NR
13	Fong et al. [2]	2009	53	–	Ancient	NR
14	Present case	2011	43	–	Conventional	NR

NF, neurofibromatosis; NR, no recurrence; UK, unknown.

encapsulation, which makes total resection possible. Complete surgical excision is the treatment of choice for vulvar neurilemmoma, the same as for those at other sites. The prognosis is excellent. The only reported case of vulvar neurilemmoma of the plexiform type recurred twice [4]. The patient was alive and well three years after the second resection. Unlike plexiform neurofibroma, which occurs almost exclusively in patients with NF1, the large majority of plexiform neurilemmoma are sporadic tumors in the vulva [1]. Malignant progression is extremely rare.

Although rare, neurilemmoma should be considered in the differential diagnosis of vulvar masses.

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소음순에 발생한 음문 신경집종 1예

가천의과학대학교 길병원 [1]병리과학교실, [2]산부인과학교실
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신경집종은 신경집에서 발생하는 양성종양으로서, 인체 어디에서나 발생할 수 있지만 여성의 음문에서 발생하는 경우는 극히 드물다. 현재까지 전 세계적으로 13예의 음문 신경집종이 문헌으로 보고되었으며 본 증례는 국내에서 처음 보고되는 음문 신경집종으로 생각된다. 43세의 여성이 좌측 소음순에 0.6×0.6 cm 크기의 결절을 주증상으로 내원하였다. 절제술을 시행하였고, 병리소견에서 고밀도의 Antoni A 영역과 성긴 Antoni B 영역이 존재하는 결절을 보였다. Antoni A 영역은 뒤틀린 핵과 핵내공포를 포함하는 방추세포로 구성되었는데, 방추세포는 짧은 다발 형태를 보였으며, Antoni B 영역은 점액성의 저세포성이었다. 면역조직화학검사에서 방추세포는 S100 단백에 양성이고 desmin 음성이었다. 빈도는 매우 낮으나, 음문 결절의 감별진단에 신경집종을 고려하여야 한다.

중심단어: 신경집종, S100 단백, 음문