

Nodular Fasciitis of the Vulva

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Abstract

Nodular fasciitis (NF) is a benign fibrous proliferative lesion, which histologically mimic a spindle cell sarcoma. It is easy to misdiagnose NF as a malignant lesion, especially in the unusual location. NF of the vulva is very rarely reported with only 13 cases on English literature. In Korea, no cases of vulvar NF have been reported. We report a case of vulvar NF occurring in a 56-year-old woman. The mass was relatively well-demarcated and measured 3.2 x 3.0 x 2.5 cm. Microscopically, the mass was composed of spindle cells with alternating hypercellular and hypocellular hyalinized fibrous areas. Mitotic figures were 1–2/10 High-Power Field. Immunohistochemical study of the mass for smooth muscle actin (SMA) yielded positive result. The CD34, desmin and ALK were negative. Ki-67 index was about 1%. Vulvar NF is noticeable to gynecologists and pathologists, because of its potential for misdiagnosis.

Key Words : Nodular fasciitis, Vulva

Introduction

Nodular fasciitis (NF) is a benign spindle cell proliferative lesion, usually involving the subcutaneous fascia of the extremities and trunk [1]. Spindle cell tumor or tumor-like lesion in the vulva is rare and of wide range, especially including leiomyoma, aggressive angiofibroma, angiofibrosarcoma, leiomyosarcoma and malignant fibrous

histiocytoma among spindle cell sarcomas. NF of the vulva is very rare. To our knowledge, 13 cases have been reported in English literature and no case in Korea [1–7]. Histologically, NF mimic spindle cell sarcoma and may be misdiagnosed as various sarcomas especially in the unusual location like vulva. We report a case of the vulvar NF to bring attention to this lesion in the vulva and to emphasize the importance of its recognition.

Clinical Presentation

A 56-year-old woman, who is in a post-menopausal state, was admitted to our clinic due to a mobile and painless mass in the left labium majora. The mass presented with a rapid growth for a month. She had not any underlying disease and didn't have a trauma history on perineal area. Gynecologic examination was unremarkable except the movable mass in subcutis. Computed tomography showed a 2.5 cm sized solid mass with well enhanced wall in subcutaneous tissue (Fig. 1). The mass was regarded as hemangioma and was completely excised under local anesthesia.

Pathologic findings

On gross examination, the mass measured 3.2 x 3.0 x 2.5 cm, and was poorly defined, round and rubbery-firm. The cut surface of the mass was gelatinous and brownish red. Necrosis and hemorrhage were absent.

Microscopically, the mass was composed of proliferation of short spindle cells with alternating hypercellular and hypocellular hyalinized fibrous areas. The spindle cells formed short bundles or fascicles and were separated by slit-like spaces. Extravasated red blood cells, lymphocytes and plasma cells infiltrates were identified. Vasculature was not well developed. The individual cells revealed mild atypia and occasionally had vesicular nuclei and prominent nucleoli. Mitotic figures were 1-2/10 High-Power Field and there was no atypical mitosis. Necrosis was not observed (Fig. 2A-C).

Immunohistochemical study of the mass for SMA yielded positive result (Fig. 2D). The

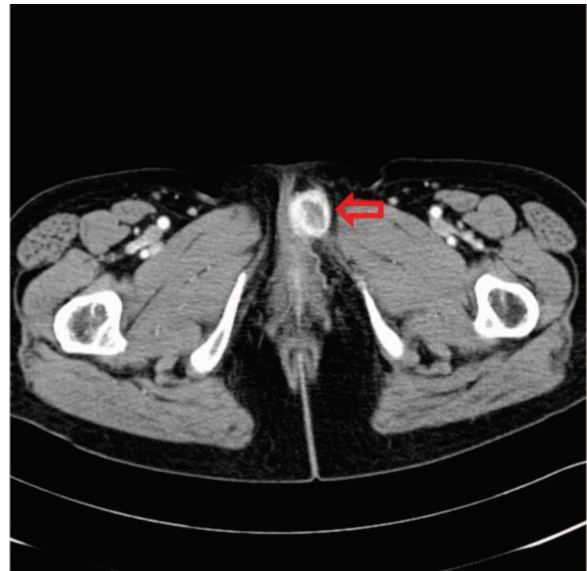


Fig. 1. Pelvic computed tomography shows a solid mass, measuring 2.5 cm in diameter in the left vulva. The mass seems to be located subcutaneous layer. The wall of the mass is strongly enhanced (arrow).

CD34, desmin and ALK stains for exclusion of smooth muscle tumor, dermatofibrosarcoma protuberans and inflammatory myofibroblastic tumor were negative. Ki-67 index was about 1%. The histologic features in conjunction with immunohistochemical stainings were consistent with NF. No evidence of recurrence was noted for 13 months follow-up.

Discussion

NF is a benign proliferative lesion of fibroblasts (or myofibroblasts). It was first described by Kornwaler *et al.* in 1955 [8]. Most cases present as a small, solitary and rapidly growing subcutaneous soft tissue mass like present case [9,10]. Usually, young and middle-aged adults under 50 years are affected. Although it may occur anywhere,

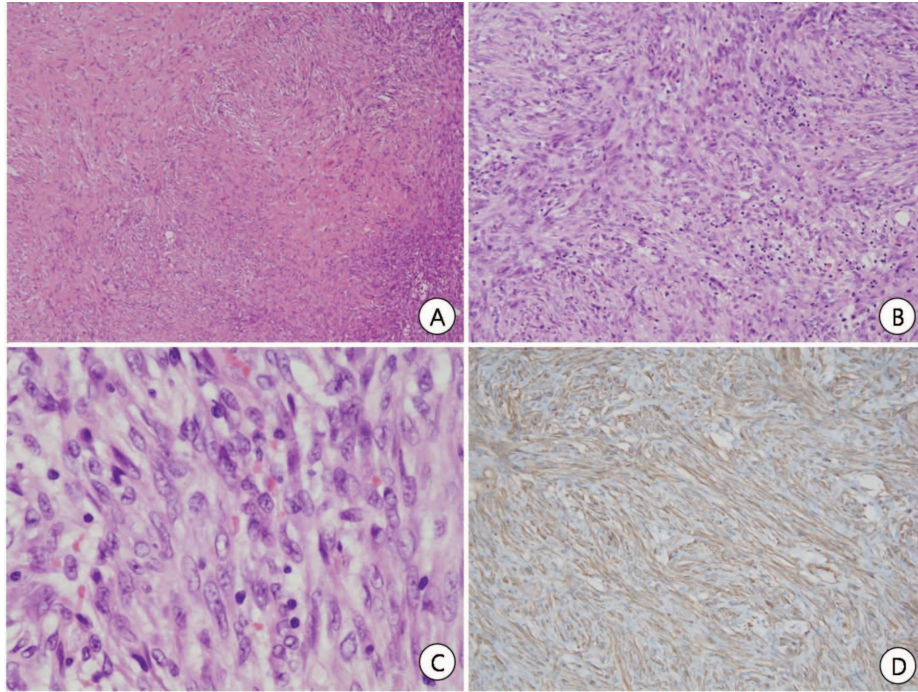


Fig. 2. Microscopic finding of the vulvar mass reveals admixture of hypercellular and hypocellular areas (A, x40). The proliferating cells are short spindle or ovoid cells formed short bundles or fascicles. They are separated by small clefts or slit-like spaces. Inflammatory cell infiltrates are present (B, x100). The spindle cells show mild atypia with occasionally vesicular nuclei and prominent nucleoli. Extravasated red blood cells are identified (C, x400). (Hematoxylin-Eosin stain) Immunohistochemistry showed spindle cells to be positive for smooth muscle actin (D, x200).

there are predilection sites. The most common site is upper extremities followed by trunk, head and neck and lower extremities [10]. The present case is very unusual in that it occurred in the vulva of relatively old-aged women. NF arising in the vulva is extremely rare with only 13 cases in English literature and no case in Korea [1–3, 5–7]. In female genital tract, NF has been reported only in the vulva except one case reported in the round ligament [11].

NF is relatively well circumscribed or occasionally poorly marginated and is not encapsulated on gross examination. Depending on amount of fibrous or myxoid stroma, it may be soft or firm and the cut surface is gelatinous

or fibrous. Occasional cystic changes may be observed. Histopathologically, the margin of NF is at least focally infiltrative. It has characteristic features, showing short broad fascicles of spindle cells of variable cellularity with small intercellular clefts and occasional cyst-like spaces. Intervening stroma is loose and myxoid or occasionally fibrous. The spindle cells are plump and considered to be fibroblasts or myofibroblasts. By immunohistochemistry, tumor cells are positive for SMA and rarely for desmin. These features are consistent with the presumptive myofibroblastic nature. The tumor cells show no frank nuclear atypia. Mitoses are seen frequently but atypical forms are absent. Associated features are

Table 1. Summary of 14 cases of vulvar nodular fasciitis

No.	Reference	Age	Site	Size (cm)	Initial Diagnosis	Follow-up
1	Present case	56	Left labium major	3.2	Nodular fasciitis	NED at 16 mo
2	Biedrzycki (7)	34	Labium major	3.0	Nodular fasciitis	NED at 5 mo
3	Aranda (6) 1998	40	Left labium major	2.0	Nodular fasciitis	NED at 6 yr
4	O'Connel (5) 1997	31	Left labium major	3.5	Atypical leiomyoma	Recurrence 4 mo after incomplete excision. Re-excision of area, NED at 6 mo
5		51	Vulva	2.2	Nodular fasciitis	NED at 3 mo
6		38	Right labium major	1.5	Angiomyofibroblastoma	NED at 2 yr
7		7	Vulva	2.5	Rhabdomyosarcoma	NED at 7 yr
8		33	Labium minus	1.5	Neurofibroma	NED at 10 mo
9		43	Left labium major	2.0	Fibroma/aggressive angiomyxoma	NED at 15 mo
10	Livolsi (4) 1987	15	Left labium major	2.5	Nodular fasciitis	NED at 1 yr
11		18	Right labium	3.0	Low-grade sarcoma of fibrous or smooth muscle origin	NED at 6 mo
12	Gaffney (1) 1982	32	Right labium	3.0	Nodular fasciitis	Re-excision of area, NED at 6 mo
13	Roberts (3) 1981	19	Right labium minor	2.5	Leiomyosarcoma	Re-excision of area, NED at 2 yr
14	Allen (2) 1972	NA	Labium major	NA	NA	NA

NA: not available, NED: no evidence of disease

variable numbers of small blood vessels, a sparse but diffuse chronic inflammatory cells infiltrate, and extravasated RBCs [1]. Multinucleated giant cells may be noted.

NF is a well known so called 'pseudosarcoma' and mimic sarcoma in that it shows rapid growth, not infrequently high cellularity and high mitotic counts. Especially in unusual location like vulva, it can be misdiagnosed as a malignancy. Since the first report of NF in the

vulva by Allen *et al.* [2], this lesion in vulva has been confused with various diseases including atypical leiomyoma, angiomyofibroblastoma, neurofibroma, fibroma, aggressive angiomyxoma, rhabdomyosarcoma, leiomyosarcoma and low-grade sarcoma (Table 1). Among 12 previous NF in vulva with known initial diagnosis, only 5 cases were initially diagnosed as NF and 3 cases were initially diagnosed as malignancy. NF is generally

smaller than 3.0 cm. It has bland nuclei and lacks atypical mitoses. A vast myofibroblasts in a stroma containing scanty collagen is a typical finding. The presence of lymphocyte infiltration, intercellular clefts, and larger cyst-like spaces are also characteristic [1]. These typical features in conjunction with appropriate immunohistochemical staining results (SMA+, CD34-, desmin-) will lead to the proper diagnosis of NF. However, several mesenchymal lesions including aggressive angiomyxoma, angiomyoibroblastoma, benign and malignant smooth muscle tumor and postoperative spindle cell nodule need to be carefully differentiated in that those lesions have predilection for vulva and show myoid trait with overlapping immunohistochemical staining result [5].

NFs are self-limited disease and cured with simple excision. They scarcely ever recur or metastasize in most cases [12]. Recurrence after incomplete excision was reported only in 1–2% of NF patients [5,12]. In vulvar NF, O'Connell *et al.* reported that only one case recurred among 6 cases of incomplete excision [5].

The awareness of distinguishing feature of NF, a benign reactive lesion from the malignant mesenchymal tumors in vulva is important to prevent overtreatment owing to its misdiagnosis and predict the prognosis.

Summary

Various mesenchymal tumors and tumor-like lesions can arise in the perineal area, including the vulva. NF of the vulva is a benign proliferative lesion. Most of the cases of vulvar NF were not initially interpreted

correctly and have been diagnosed as various other disease entities, both benign and malignant. Vulvar NF must be recognized to avoid misdiagnosis and overtreatment.

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