Dermatofibrosarcoma protuberans of the mons pubis

A. Zizi-Sermpetzoglou[1], V. Savvaidou[1], S. Fournogerakis[2], E. Moustou[1], M. Konstantidelli[2], N. Vlachakos[2]

[1]Department of Pathology, [2]Second Department of Surgery, Tzaneion General Hospital of Piraeus, Piraeus (Greece)

Summary

Dermatofibrosarcoma protuberans (DFSP) is a rare fibrous tumor with intermediate malignant potential and in rare cases the vulva is involved. The most common clinical presentation is a firm plaque with surrounding red to blue discoloration, or less often with multiple small subcutaneous nodules. The authors present a case of a 66-year-old woman who came to the hospital complaining of longstanding painless nodules in the area of the mons pubis. On physical examination, a diffuse area of erythematous induration involving the mons pubis was recognized and within this area there were smaller nodules. The histological diagnosis was dermatofibrosarcoma protuberans. Microscopically DFSP has a storiform pattern of uniform cytologically bland spindle cells, with a characteristic honeycomb pattern of infiltration into the subcutaneous fat. Immunohistochemical staining demonstrates strong positivity for vimentin and CD34. The treatment has been through a wide local excision (WLE), although microscopic tumor projections beyond the central tumor nodule explain the tumors propensity for local recurrence.

Key words: Vulva; Dermatofibrosarcoma protuberans; Immunohistochemistry; Treatment.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous tumor of intermediate grade malignancy, with a tendency for local recurrence [1, 2]. It rarely occurs in the vulva and in a review of the literature, less than 35 cases of DFSP in this area have been reported [3, 4]. Preferred treatment for DFSP is wide surgical excision with pathologically negative margins [5]. Despite the local invasiveness it rarely metastasizes [6].

The authors present another case of DFSP arising from the mons pubis and discuss its clinical presentations, histological characteristics, treatment modalities, and outcomes.

Case Report

A 66-year-old woman was admitted to the Hospital complaining of a long-standing painless nodule in the area of mons pubis. She had a history of breast cancer 15 years prior treated with surgery, but neither chemotherapy nor radiotherapy (RT) was given. On physical examination at the time of presentation, there was a diffuse area of erythematous induration involving the mons pubis measuring 8 x 5 cm. Within this area, there were smaller nodules. The rest of the gynecologic examination was normal. There was no lymphadenopathy or other skin abnormalities. The patient was offered vertical excision with the reconstruction of the area. The specimen was an vulvar ellipse measuring 8 x 5 cm. On gross examination, subcutaneously three nodules of various sizes (1, 2, and 2.5 cm) were found. The nodules were hard in consistency with a light brown color. Two more nodules were found in the adipose tissue. They were also hard in consistency, of pale white color and multilobulated. Microscopically, in both the dermis and subcutaneous fat a monotonous storiform pattern of uniform, bland spindle cells is recognized (Figures 1 and 2). There is little nuclear pleomorphism. The number of mitoses is less than five/ten high power fields (hpf). In some areas the myxoid change of the tumor is prominent. Immunohistochemically the tumor cells were diffusely and strongly positive for vimentin (Figure 3), and CD34 (Figure 4) and negative for smooth muscle actin, CD117, S100P, progesteron, and oestrogen.

On the basis of histological and immunohistochemical findings, the diagnosis of DFSP of mons pubis was established. The patient underwent further diagnostic procedure consisting of ultrasound scan of the thorax without evidence of the disease in any other organ or lymph node tissue.

Discussion

DFSP, first described in 1924 by Darrier and Ferrand [7], is a nodular cutaneous tumor characterized by a prominent storiform pattern. These tumors occur at any site, but they are seen most frequently in the trunk and proximal extremities. DFSP of the female genital area, such as the vulva, is extremely rare. In a review of the literature, less than 35 cases of vulvar DFSP have been reported [3, 4]. DFSP of the vulva accounts for 0.1% of all malignancies [8]. Patient age ranged from 23 to 76 years (mean age 46). Vulvar DFSP may present as a firm plaque of the skin with subcutaneous nodules with surrounding red to blue discoloration or as multiple small subcutaneous nodules, such as in the presented case. Histologically DFSP has a more distinct storiform pattern, little nuclear pleomorphism, and low to moderate mitotic activity. Necroses are absent [8, 9]. Immunohistochemically, DFSP is characterized by the presence of CD34 [10, 11]. Ultrastructurally earlier studies showed that the cells resemble fibroblasts, but the expression of CD34 (human progenitor cell antigen) supports the view that this neoplasm is a variant of nerve sheath tumor. Also, it is characterized by a chromosomal translocation between distinct regions of chromosomes 17 and 22, t (17; 22)

Revised manuscript accepted for publication July 13, 2011

Fig. 1

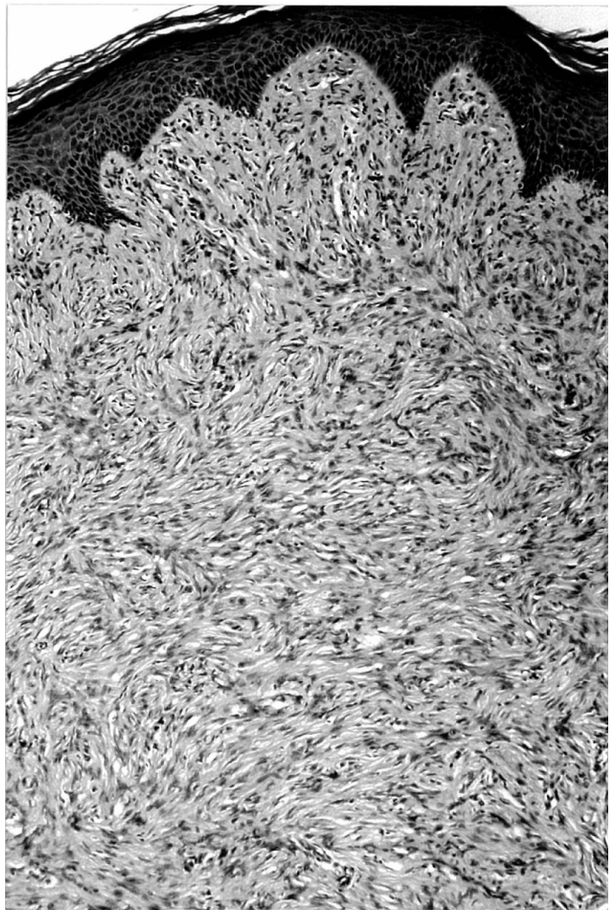

Fig. 2

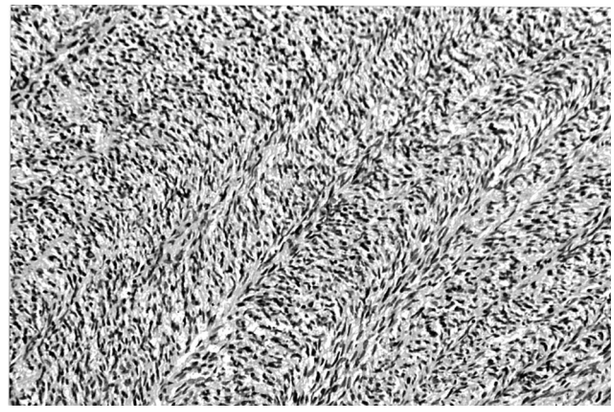

Fig. 3

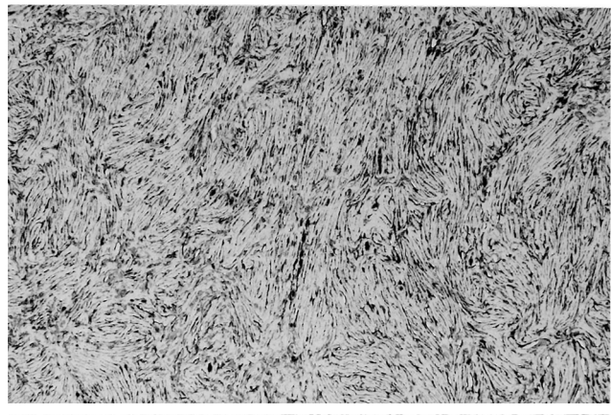

Fig. 4

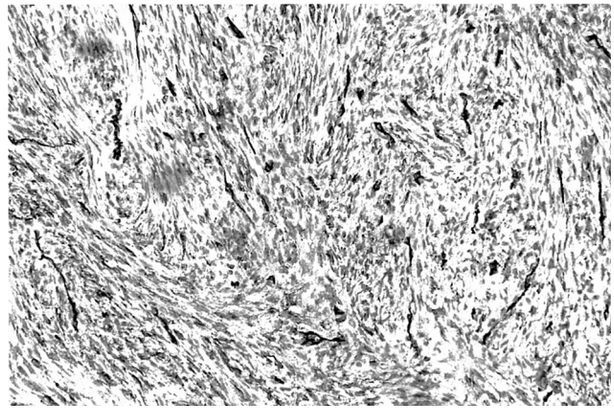

Figure 1. — Dermal tumor composed of spindle cells that extend to the subcutis (H&E x 100).
Figure 2. — DFSP whorled pattern (H&E x 200).
Figure 3. — DFSP spindle cells strongly positive for vimentin (vimentin x 200).
Figure 4. — DFSP spindle cells strongly positive for CD34 (CD34 x 200).

leading to fusion of the collagen type 1 alpha1 gene (COLIA1) to the platelet-derived growth factor B (PDGFB) gene [12]. Overproduction of PDGFB by DFSP results in autocrine stimulation. In the differential diagnosis of DFSP, other soft tissue tumors must be affected such as dermatofibrosarcoma with its variants, neurofibroma, shwannoma, malignant peripheral nerve sheath tumors (MPNST), leiomyoma, leiomyosarcoma, myxoid liposarcoma, desmoplastic melanoma, and solitary fibrous tumor. The final diagnosis is made with the help of positivity for CD34.

The treatment of DFSP has been with a wide local excision (WLE). The local recurrence rate after WLE ranges from 20-50%, because there are microscopic tumor projections beyond the central tumor nodule resulting in incomplete excision [5]. Mohs micrographic surgery (MMS) is an effective treatment of DFPS. MMS allows mapping the extension of DFSP with microscopic examination of its deep and lateral margins and enables to examine 100% of the tumor margin [5, 13]. MMS used to treat DFSP of the trunk and extremities is well-documented [13]. The local recurrence rate after the MMS treatment for patients with DFSP has been reported to be 1.6% vs 20% after wide excision and 43% after conservative excision [14].

In the vulva, the treatment of DFSP ranges from simple excision to radical vulvectomy and RT [15]. Until recently treating vulvar DFSP with MMS was referred in

a limited number of cases. In these cases the tumor did not recur, while the recurrence rate in patients treated with non-MMS modalities was 32% [16, 17]. This method requires trained staff and experienced surgeons. Finally, imaging studies such as computed tomography (CT) scans, magnetic resonance imaging (MRI), and X-ray of the thorax are necessary to determine the extent of tumor size because finger-like projections into surrounding tissue increase dangerous recurrences.

In conclusion, vulvar DFSP is an uncommon tumor, with less of 35 cases, that has the tendency to recur locally. The diagnosis is established with the help of immunohistochemistry [CD34 (+)]. Most of the cases are characterized by a reciprocal translocationt t (17; 22) (q22; q13).

The treatment of vulvar DFSP is with a wide simple excision. More recently MMS is an effective treatment, but this method requires trained staff and experienced surgeons.

References

[1] Mentzel T., Beham A., Katenkamp D., Dei Tos AP, Fletcher C.D.: "Fibrosarcomatous "high grade" dermatofibrosarcoma protuberans: clinicopathologic and immunohistochemical study of a series of 41 cases with emphasis on prognostic significance". *Am. J. Surg. Pathol*., 1998, *22,* 576.

[2] McArthur G.A.: "Dermatofibrosarcoma protuberans: a surgical disease with a molecular savior". *Curr. Opin. Oncol.,* 2006, *18,* 341.

[3] Oge T., Benedicic Ch., Tamussino K., Regauer S.: "Dermatofibrosarcoma protuberans of the vulva: a case report". *BMJ Case Reports*, 2009 doi: 10.1136/bcc.07.2008.0377.

[4] Edelweiss M., Malpica A.: "Dermatofibrosarcoma Protuberans of the vulva. A clinicopathologic and immunohistochemical study of 13 cases". *Am. J. Surg. Pathol*., 2010, *34,* 393.

[5] Gloster H.M. Jr., Harris K.R., Roenigk R.K.: "A comparison between Mohs micrographic surgery and wide surgical excision for the treatment of dermatofibrosarcoma protuberans". *J. Am. Acad. Dermatol*., 1996, *35,* 82.

[6] Soergel T.M., Doering D.L., O'Connor D.: "Metastatic dermatofibrosarcoma protuberans of the vulva". *Gynecol. Oncol.,* 1998, *71,* 320.

[7] Darrier J., Ferrand M.: "Dermatofibromes progressives at recidivants ou fibrosarcomas de la peua". *Ann. Dermatol. Syphilgr*. (Paris), 1924, *5,* 545.

[8] Olejek A., Kozak-Darmas I., Gajewska A., Gabriel A., Zajecki W.: "Dermatofibrosarcoma protuberans of the vulva". *Wiad Lek.,* 2008, *61,* 232.

[9] Moodley M., Moodley J.: "Dermatofibrosarcoma protuberans of the vulva: a case report and review of the literature". *Gynecol. Oncol.,* 2000, *78,* 74.

[10] Ghorbani R.P., Malpica A., Ayala A.G.: "Dermatofibrosarcoma protuberans of the vulva: clinicopathologic and immunohistochemical analysis of four cases, one with fibrosarcomatous change and review of the literature". *Int. J. Gynecol. Pathol.,* 1999, *18,* 366.

[11] Kutzner H.: "Expression of human progenitor cell antigen (CD34 (HPCA-1) distinguishes dermatofibrosarcoma protuberans from fibrous histiocytoma in formalin-fixed,paraffine embedded tissue". *J. Am. Acad. Dermatol*., 1993, *28,* 613.

[12] Gokden N., Dehner L.P., Zhu X., Pfeifer J.D.: "Dermatofibrosarcoma protuberans of the vulva and groin: detection of COL 1A1-PDGFB fusion transcripts by RT-PCR". *J. Cutan Pathol.,* 2003, *30,* 190.

[13] Wacker J., Khan-Durani B., Hartschuch W.: "Modified micrographic surgery in the therapy of dermatofibrosarcoma protuberans: analysis of 22 patients". *Ann. Surg. Oncol.,* 2004, *11,* 438.

[14] Snow S.N., Gordon E.M., Larson P.O.: "Dermatofibrosarcoma protuberans: a report of 29 patients treated by mohs micrographic surgery with long term follow-up and review of the literature". *Cancer*, 2004, *101,* 28.

[15] Ballo M.T., Zagars G.K., Pisters P., Pollack A.: "The role of radiation in the management of dermatofibrosarcoma protuberans". *Int. J. Radiat. Oncol. Biol. Phys*., 1998, *40,* 823.

[16] Doufekas K., Duncan T.J.,Williamson K.M., Varma S., Nunns D.: "Mohs micrographic surgery for dermatofibrosarcoma protuberans of the vulva". *Obstet. Gynecol*., 2009, *54*, 7672.

[17] Hammonds L.M., Hendi A.: "Dermatofibrosarcoma protuberans of the vulva treated using mohs micrographic surgery". *Dermatol. Surg*., 2010, *36*, 558.

Address reprint requests to:
A. ZIZI-SERMPETZOGLOU, M.D., Ph.D.
P.O. Box 3143
Alikos
19400 Ag. Marina, Koropi
Athens (Greece)
e-mail: adserbet@yahoo.gr