Genital Desmoplastic Fibroblastoma (Collagenous Fibroma)

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INTRODUCTION

Desmoplastic fibroblastomas (DFs) are rare fibrous soft tissue tumors that usually arise in subcutaneous tissue or skeletal muscle in a variety of anatomical sites. This was first described by Evans in 1995 and was classified as a distinctive form of benign fibrous soft tumor.[1] In 1996 the lesion was renamed as a " collagenous fibroma" by Nielsen and colleagues.[2] The arm or the shoulders are the most frequent sites of involvement. They have also been described in the neck, tongue, lacrimal gland and palate.[3-7] To the best of our knowledge, we report the first case of DF (collagenous fibroma) occurring in genital area.

CASE REPORT

A 71-year old man presented with a giant multiple globular mass in the scrotum which has grown slowly for

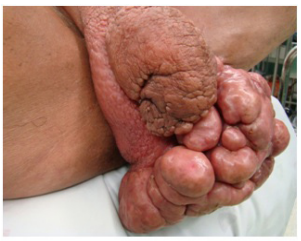

Figure 1. The 20 × 15 cm sized, homogeneous pearl-grey colored, firm globular mass in the scrotum has extended to adjacent penis and left inner thigh. Penile glans was not appreciable.

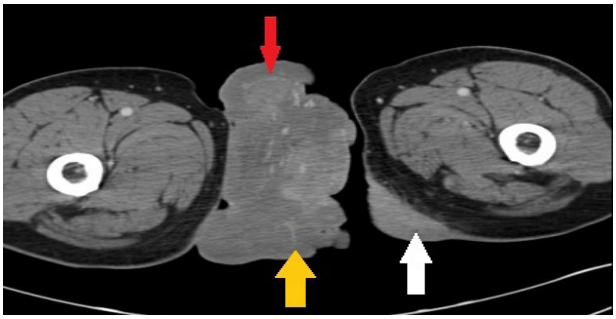

Figure 2. Computed tomography scan showed a 15 × 9 cm sized soft tissue mass in the scrotum (yellow arrow) and penis which has extended into the fascial area of left inner thigh (white arrows). Penile glans (red arrow) is trapped in within the mass.

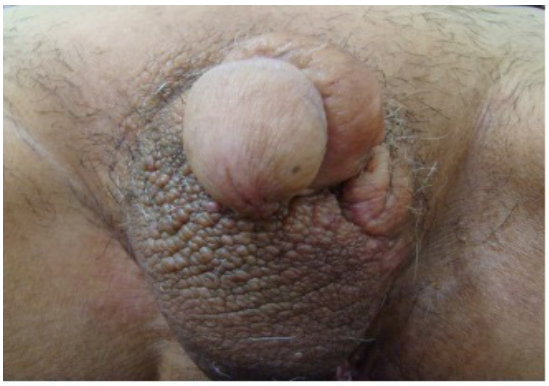

Figure 3. Postoperative finding after excision of mass. Penile glans is exposed.

4 years. On physical examination, a giant multiple globular mass in the scrotum has extended to adjacent penis and left inner thigh. Penile glans was not appreciable (**Figure 1**). He had no history of genital surgery. A pelvic computed tomography (CT) scan showed a soft tissue mass in the scrotum and penis which has extended into the fascial area of left inner thigh (**Figure 2**). The patient underwent total excision of the mass in the scrotum, penis and left inner thigh. Skin defect in penile shaft and scrotum were managed with penile skin graft and scroto-plasty (**Figure 3**). On pathology, the resected penile and scrotal masses measures 17.5 × 11 cm and 12 × 8 cm, respectively, with vaguely circumscribed subcutaneous lesion with convoluted skin surface. The cut surface was whitish grey in color without hemorrhage or necrosis. Microscopically, the mass was paucicellular and consists of widely spaced bland spindle- to stellate-shaped fibroblasts embedded in a collagenous and myxocollagenous stroma (**Figure 4**). Cellular atypia or abnormal mitosis was absent. The immunohistochemical stains showed vimentin positive in stellated fibroblast, but, desmin, smooth muscle actin, S-100 protein, CD34, CD68, factor-8, myoglobin and neurofilament were negative. Tumor recurrence was not observed for 12 months.

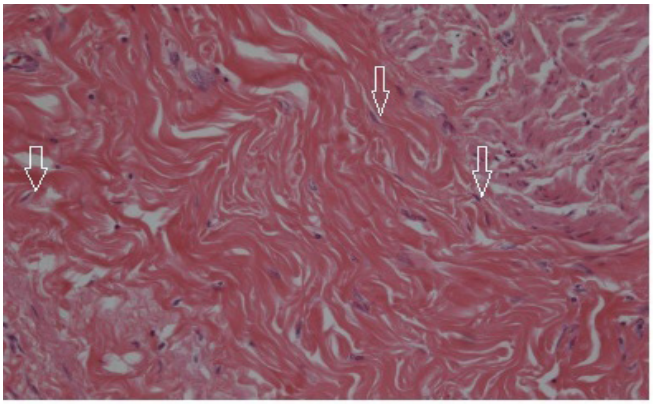

Figure 4. Microscopically, the tumor has been composed of hypocellular spindle to stellate shaped fibroblasts and myofibroblasts embedded in a prominent collagenous stroma (white arrows) (Hematoxylin and eosin stains × 100).

DISCUSSION

DFs also known as collagenous fibromas are benign soft tissue paucicellular tumors. They are usually well circumscribed and are composed of spindle to stellate shaped fibroblasts dispersed in a fibromyxoid or densely fibrous background stroma with low mitotic activity. So, Miettinen and Fetsch recommended the designation stellate cell fibroma. Immunohistochemical and ultrastructural studies show that the tumor cells are predominantly fibroblastic in nature and typically positive for vimentin.[3]

There is often focal reactivity for muscle actins (HHF-35) and α-smooth muscle actin.[3] Scattered CD68-positive histiocytes and mast cells may be present, but the tumor cells are negative. There is no documented immunoreactivity for CD34, S-100 protein, desmin, or epithelial membrane antigen (EMA). The lesion typically presents with a long history of a painless, slowly growing well-circumscribed subcutaneous mass occurring predominantly in males, with a median age of 50 years.[1,2] Since 1995, approximately 94 cases of DF have been reported in the literature with the largest case series of 63 patients being published by Miettinen and Fetsch.[3] It appears in a variety of peripheral sites with the most common location being the arm, shoulder, lower limb, back, forearm, hands, feet, neck and even in the tongue, lacrimal gland , palate and parotid gland.[3-8]

CONCLUSION

In conclusion, the present case is the first description of a DF that has involved the genital area. The clinical, gross and histologic features are those of a benign neoplasm. We highlight this peculiar lesion and wish to increase awareness of these rare lesions among urologists and pathologists alike.

CONFLICT OF INTEREST

None declared.

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