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Case Report

Myxoid dermatofibrosarcoma protruberans of the scrotum: A rare diagnosis in an unreported location

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Abstract Dermatofibrosarcoma Protuberans (DFSP) represents a low grade, locally aggressive mesenchymal neoplasm with characteristic clinicopathologic, immunohistochemical, and molecular findings. Myxoid DFSP is an uncommon variant, with only few cases reported in the literature and may present a diagnostic challenge on histopathologic examination. We report the first case of myxoid DFSP arising in the scrotum of a 38 year-old man and describe the morphologic and immunohistochemical findings. Recognition of this variant at unusual sites is clinically important because the differential diagnosis includes benign and malignant tumors which could lead to under or over-treatment.

1. Introduction

Dermatofibrosarcoma Protruberans (DFSP) is a superficial, low grade locally aggressive mesenchymal neoplasm of fibroblastic origin which arises in the dermis and frequently involves the subcutaneous tissues. It arises most commonly in the trunk, extremities, and head and neck but may involve any body site [1]. Although it can occur at any age, it typically presents in young to middle-aged adults, with a peak incidence in the fourth decade of life. Cytogenetically, DFSP is characterized by an identical t(17;22)(q22;q13) translocation, with some cases resulting in a ring chromosome, forming a chimeric COL1A1-PDGB gene [2], which plays an important role in DFSP pathogenesis [3]. The typical histologic feature of DFSP is a storiform proliferation of bland spindle shaped cells infiltrating the subcutaneous fat in a honeycomb pattern, however a wide morphologic spectrum can be observed. Several histologic variants are recognized including pigmented DFSP (Bednar tumor), fibrosarcomatous DFSP, myxoid DFSP, flat atrophic DFSP, giant cell fibroblastoma, and DFSP with myogenic differentiation [4–8].

Myxoid DFSP represents a rare morphologic variant with prominent myxoid stromal changes that may cause considerable diagnostic challenges, particularly in the distinction from other more clinically aggressive myxoid mesenchymal neoplasms. To the best of our knowledge, this is the first case of myxoid DFSP involving the scrotum. We describe the morphologic and immunohistochemical findings and discuss the differential diagnosis of this rare variant.

2. Materials and methods

2.1. Case report

A 38 year-old man with a long-standing history of perineal hidradenitis, presented to the Urology clinic complaining of an

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enlarging, painless, left scrotal mass that was originally the size of a golf ball and had gradually increased in size over a period of two years. As part of the workup, an ultrasound of the pelvis was performed which showed a mass in the left paratesticular area. A decision was made to surgically excise the mass.

Intraoperatively, two left scrotal masses, discrete from the testes, were identified and excised, each measuring 5.0 cm in greatest dimension. An area of hidradenitis along the left hemiscrotum, measuring 7.0 cm in greatest dimension was also identified and resected.

2.2. Histology and immunohistochemistry

Tissue sections were fixed in 10% neutral buffered formalin, embedded in paraffin, sectioned at 4-μm thickness, and stained with hematoxylin and eosin. The immunohistochemistry was performed and evaluated at Wayne State University (Detroit Medical Center, Detroit, MI), where the Ventana BenchMark Autostainer (Ventana Medical System, Tucson, Arizona) was used on 4-μm thick formalin-fixed and deparaffinized sections with the following markers: CD34, smooth muscle actin, AE1/AE3, desmin, S100, CD31 and MUC4.

3. Results

3.1. Gross and microscopy

Grossly, the specimen consisted of tan–yellow soft tissue, partially covered by skin that was remarkable for four raised nodular areas ranging in size from 0.6 cm up to 1.5 cm. Sectioning revealed two pink–gray, well circumscribed fleshy masses measuring 5.5 × 4.8 × 3.0 cm and 5.0 × 4.5 × 3.7 cm that appeared to be connected resulting in a dumbbell-shaped mass with an overall measurement of 10.5 × 4.8 × 3.7 cm. Cut surfaces were gelatinous and tan–gray. Histologic examination revealed an ill-defined lesion with an infiltrative pattern of growth, involving the dermis and extending into the subcutaneous tissues and present at the resection margin. The lesion was variable in cellularity with relatively hypocellular areas with myxoid stroma and numerous thin-walled blood vessels, alternating with hypercellular areas forming a storiform pattern (Fig. 1). Cytologically, the cells were low grade, spindle shaped, relatively uniform with oval hyperchromatic nuclei and inconspicuous nucleoli. Occasional mitotic figures were seen. Myoid balls were identified surrounding blood vessels (Fig. 2). In some sections, the overlying skin was remarkable for features of Hidradenitis suppurativa including heavy neutrophilic and mixed inflammatory infiltrate around apocrine glands as well as sinus tracts with marked acute inflammation, abscess formation and occasionally lined by granulation tissue.

3.2. Immunohistochemistry

The tumor cells were diffusely positive for CD34 and smooth muscle actin but were negative for cytokeratin, ALK-1, desmin, S100, CD31 and MUC4 immunostains (Table 1).

Based on the morphology and immunophenotype, a diagnosis of myxoid DFSP was rendered. The patient is now few months post surgery with no evidence of disease.

4. Discussion

Myxoid DFSP was first described by Freirson and Cooper in 1983 [8], who reported two cases of myxoid DFSP that

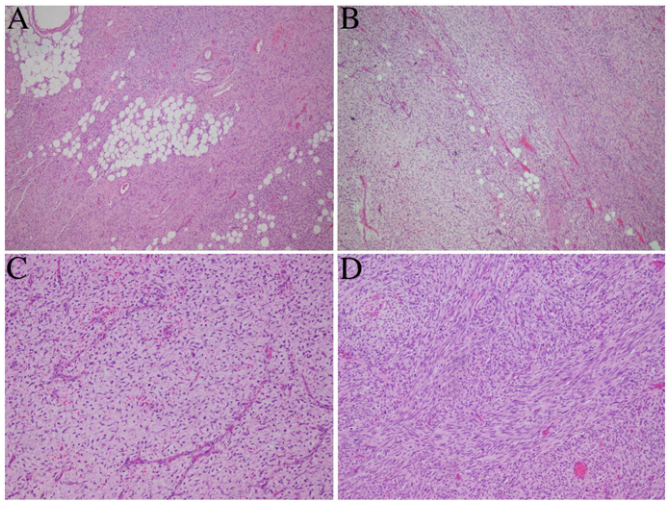

Fig. 1 A. The tumor extends from dermis into subcutaneous adipose tissue with trapping of fat in a "honeycomb" pattern (40×). B. The tumor consists of a relatively hypocellular myxoid areas alternating with more cellular areas (40×). C. Delicate, thin-walled vessels were frequently present within the myxoid stroma (100×). D. The relatively hypercellular area consisting of spindle-shaped cells forming a storiform pattern (100×).