Proliferating Trichilemmal Tumor Presenting as a Scrotal Mass in a Middle-Aged Male: An Uncommon Location

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Abstract

Scrotal masses encompass a heterogeneous group of conditions affecting the skin and soft tissue of the scrotum or underlying testis, ranging from benign to malignant and from indolent to conditions requiring urgent medical evaluation. Accurate and timely diagnosis is imperative with scrotal masses, as the medical ramifications of different diagnoses vary widely, and the social and psychological implications can be significant. Here we present the first reported case of a middle-aged male with an enlarging scrotal/extra-testicular mass, which upon excision, was found to be consistent with a proliferating trichilemmal tumor, a benign neoplasm derived from outer root sheath of the hair follicle which has a strong capacity to simulate squamous cell carcinoma. Distinguishing between these two entities is of utmost importance due to their significantly different prognostic and treatment ramifications, and ultimately directs appropriate management of these patients.

Keywords: Scrotum; Testicle; Testis; Testes; Squamous cell carcinoma; Male; Proliferating; Trichilemmal tumor; Neoplasm

Introduction

Scrotal masses encompass a heterogeneous group of conditions affecting the skin and soft tissue of the scrotum or underlying testis, ranging from benign to malignant and from indolent to conditions requiring urgent medical evaluation [1]. Accurate and timely diagnosis is imperative with scrotal masses, as the medical ramifications of different diagnoses vary widely, and the social and psychological implications can be significant. Benign conditions affecting the scrotum and testes include, but are not limited to, epidermal cyst, inguinal hernia, epididymitis, orchitis, spermatocele, hydrocele, varicocele, hematocoele, and testicular torsion [1]. On the opposite end of the spectrum are malignant tumors of the scrotum and testes. While a variety of seminomatous and non-seminomatous germ cell tumors are the most common malignancies in the testes, squamous cell carcinoma (SCC) remains the most common malignancy of the scrotum [2]. SCC of the scrotum has been a topic of interest for both clinical and historical reasons. It was documented by Pott in 1775 as one of the first occupational diseases occurring as a result of chronic soot exposure in the rugal folds of chimney sweeps [3-5]. While now rare due to improved working conditions, SCC still remains a risk in a variety of industrial occupations such as car and airplane manufacturing / repair, gas distribution, engineering, textile milling and metalworking [4,6-8]. Although occupational SCC is likely due to exposure to carcinogenic polycyclic aromatic
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Figure 2: H&E, 20x. Low power view shows a well-circumscribed dermal neoplasm with no connection to the overlying epidermis and a pushing boarder.

Figure 3: H&E, 200x. High power view shows a portion of the cyst wall, lined by stratified squamous epithelium devoid of a granular layer with peripheral palisading and trichilemmal-type keratinization.
Figure 4: H&E, 200x. High power view shows the solid component, which consists of interanastomosing lobules of squamous epithelium devoid of a granular layer with foci of trichilemmal-type keratinization.

Figure 5: H&E, 400x. High power view shows no significant cytologic atypia, necrosis, atypical mitoses, or infiltration into the surrounding tissue.
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Discussion

Proliferating trichilemmal tumors (PTT) are uncommon and usually benign neoplasms arising from the isthmus region of the outer sheath of the roots of hair follicles. Wilson-Jones first described PTT in 1966 as an entity that can clinically and histologically simulate squamous cell carcinoma. Initially classified as a pseudoepitheliomatous hyperplasia, a number of other terms such as proliferating epidermoid cyst, pilar tumor of the scalp, proliferating trichilemmal cyst, proliferating epidermoid cyst, giant hair matrix tumor, hydatidiform keratinous cyst, and invasive hair matrix tumor have been used. PTT typically presents as a solitary, painless nodule, ranging in size from 1 to 10 cm. Although the majority (90%) present on the scalp, other hair-bearing anatomic sites, including the trunk, and rarely the extremities, nose, eyelid and vulva, can also be involved. There is a marked female predominance. They are characterized histologically by features of a typical pilar cyst, but additionally show extensive epithelial proliferation with a pushing boarder, mild cytologic atypia and mitotic activity. Rarely, malignant transformation can occur, which demonstrates severe nuclear atypia, atypical mitoses, necrosis and an infiltrative growth pattern. Surgical excision with a minimal negative margin is the treatment for PTT. To our knowledge, this is the first reported case of a proliferating pilar cyst presenting in the scrotum.

In contrast, squamous cell carcinoma (SCC), although rare, remains the most common scrotal malignancy with a propensity for recurrence and metastasis. It most commonly presents as a solitary erythematous nodule or plaque with or without keratinization or ulceration, most commonly on the left scrotum. Histologically, squamous cell carcinoma consists of cords, strands and islands of squamous cells originating from the overlying epidermis and invades into the dermis with an infiltrative boarder. There is significant cellular atypia, numerous atypical and typical mitoses, and variable necrosis. The cells have abundant eosinophilic cytoplasm, large vesicular and pleomorphic nuclei and prominent nucleoli. Keratin pearls can be present throughout the tumor depending on its level of differentiation. Although surgical excision is also the primary treatment for SCC, current guidelines require a wider margin (range 4 mm - 3 cm) than PTT. Additionally, due to the fact that the raphe of the scrotum does not provide a physical barrier to scrotal lymphatic drainage and that the scrotum has bilateral inguinal drainage, secondary treatments such as (1) inguinal sentinel lymph node dissection (ILND) / subsequent complete ILND in cases with clinical or histologic evidence for regional spread of disease and (2) palliative chemotherapy for locally advanced and metastatic disease are advocated for SCC. It is important for clinicians to be aware of PTT as an entity when discussing the differential diagnosis of scrotal masses, as it can be clinically and histologically confused with the more common SCC of the scrotum and has significantly different prognostic and treatment ramifications.

Conclusion

PTTs are uncommon, slow-growing tumors that can pose a diagnostic dilemma due to their ability to mimic squamous cell carcinoma, especially when presenting in an unusual location. To our knowledge, this is the first case of a PTT described in the scrotum.

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