Sarcomatoid carcinoma of the Penis: A case report and review of literature

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Abstract

Sarcomatoid carcinomas are biphasic tumors which can occur at any site in the human body. In the medical literature, very few cases have been reported as arising from the penis. A few studies consider these tumors as a variant of squamous cell carcinoma or a metaplastic differentiation of the mesenchyme. Their clinical behavior is aggressive with both blood borne and lymphatic metastases. Treatment is by surgical excision of tumor. Dissection of lymph nodes have shown both epithelial and sarcomatous components. We report a 71-year-old male with a sarcomatoid carcinoma of the penis which was confirmed by immunohistochemistry. The rarity of this entity makes it a clinicopathologic curiosity.

Discussion

Histological examination of the mass presented a malignant spindle cell tumor diagnosed by immunohistochemistry as most consistent with sarcomatoid squamous cell carcinoma. Synovial sarcoma could not be entirely ruled-out. Nevertheless, we decided to send the case to NIH for a second opinion and they concurred with the diagnosis of a sarcomatoid carcinoma of the penis.

Penile carcinoma is an uncommon malignant disease, with an incidence of 0.1-0.9 per 100,000 males in Europe and 0.7-0.9 per 100,000 males in the USA(1,8,6). However, in some areas of Asia, Africa and South America, the incidence of penile carcinoma is significantly higher at 19 per 100,000 males and accounts for as much as 10-20% of male cancers(2). Almost 95% of all penile cancers are squamous cell carcinoma (SCC)(6). The World Health Organization(WHO) classify them as usual, basaloid, verrucous, warty(condylomatous), papillary, sarcomatoid, adenosquamous and mixed(2,7). Mean age is 60-62 years(2,6). Risk factors for the development of penile carcinomas, such as phimosis, chronic conditions, absence of circumcision, and human immunodeficiency virus infection(HIV), have been identified. Human papillomavirus infection(HPV) detection in penile carcinomas previously thought to almost always occurred in basosolid and warty subtypes, sometimes in the verrucous and usual type SCC subtypes, and not documented in sarcomatoid carcinoma variant.(8) Poblet et al. reported 2 cases of penile sarcomatoid carcinoma positive to HPV by PCR (genotypes 16, 18,1,7).

Sarcomatoid SCC of the penis is a rare variant of penile cancer, representing1% to 2% of all penile carcinomas(8). It is a tumor that originates from epithelial (tumor) cell, often retaining epithelial markers on immunohistochemistry. Localization of the primary tumor appears in the glans in 48% of cases, prepuce in 21%, coronal sulcus in 6% and the shaft in less than 2%(4,9,10). Of these patients, 17-45% actually have lymphogenic and hematogenous spread(4,10,12). Of note, 75% of patients with sarcomatoid carcinoma died, usually within a year(10,12). Grossly, they are 5-7cm irregular, light grey-white, mixed exophytic and endophytic masses(1,7). Histologically, there are atypical spindle cells with features similar to fibrosarcoma, leiomyosarcoma, melanoma and malignant fibrous histiocytoma(4,8,12). These cells have the potential to differentiate into muscle, bone and cartilage, benign or malignant(10). Given the strong predictive value of lymph node involvement for overall survival , evaluating regional LNs is critical. Advanced LN involvement is increasingly being treated with multimodality therapy incorporating chemotherapy and/or radiation. A single superior cisplastin-based regimen has not been defined. Further advance may occur with a better collaboration on an international scale and comprehensive understanding of tumor biology. To this end, the preventive role of circumcision and understanding of the oncogenic roles of Human Papilloma Virus and smoking may hold advances. Preliminary data suggest a role for agents targeting epidermal growth factor receptor and angiogenesis(5,6). In nonmetastatic cases adjuvant chemotherapy should be considered to explore its effect on recurrence(10).

Microscopic Findings

At low magnification (4 X), hematoxylin & cosin stained slides of the primary tumor revealed a biphasic histopathologic pattern with areas of pleomorphic spindle cells admixed with epithelioid squamoid cells (Figure1). Higher magnification showed anaplastic tumors with a spindle and angiomatoid pattern of growth with marked mitotic activity (Figure2). Immunostain for Ki67, a proliferation nuclear marker, was positive in up to 90% of cells in some areas (Figure3). Cells were also positive to cytokeratins 8/18, 5/6 in occasional cells (Figure 4,5), while negative for CD34, desmin, SMA, and S-100.

Gross Findings

The specimen consisted of a partial penectomy. It was not circumcised and the glans was distorted by a light tan exophytic mass with necrotic areas measuring 7.5 x 7.5 x 5 cm which extended up to 0.5 cm from the margin of resection. The tumor grossly invaded the corpus spongiosum and corpora cavernosa. The urethra was obstructed by the tumor.

References


Figure 1. Disorderly biphasic pattern with areas of pleomorphic spindle cells admixed with epithelioid squamoid cells (H&E; X100)

Figure 2. Epithelioid squamoid cells admixed with pleomorphic bipolar cells with atypical nuclei (H&E; X400)

Figure 3. Ki67 positivity in more than 90% of cells (Nuclear staining; X200)

Figure 4. Cytokeratin 8/18 positive in occasional cells (Arrow. X200)

Figure 5. Cytokeratin 8/6 positive in occasional cells (Arrow. X200)