Primary Undifferentiated Pleomorphic Sarcoma of the Penis

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Background: Primary penile sarcoma is a rare disease that affects men of all ages. Different subtypes of primary penile sarcoma exist, with the rarest being pleomorphic sarcoma. Delays in presentation and diagnosis of primary penile sarcoma have been reported because of its benign-appearing presenting features and rarity. If penile sarcoma is left untreated, the clinical consequence is metastasis that is fatal in most cases.

Case Report: We report an extremely rare case of undifferentiated pleomorphic sarcoma of the penis in a 59-year-old patient who initially presented with a slow-growing penile nodule. The tumor was surgically excised, but the patient experienced local recurrence and, despite receiving chemotherapy and surgery, died of metastatic disease 15 months after initial presentation.

Conclusion: Vigilance regarding biopsy and intervention for penile nodules may lead to early diagnosis and improved clinical outcomes.

Keywords: Penile neoplasms, penis, sarcoma

INTRODUCTION

Penile cancer is uncommon and mostly consists of carcinoma, with squamous cell being the most common type. Primary penile sarcoma is a rare soft tissue tumor with an incidence of 0.6-1.0 cases per 100,000 population in developed countries (data from 1984). Common subtypes of primary penile sarcoma are Kaposi sarcoma, angiosarcoma, leiomyosarcoma, and epithelioid sarcoma. Because of its rarity, a diagnosis of penile sarcoma can be difficult to establish without an obvious presentation. Furthermore, primary penile sarcoma is often misdiagnosed because of its slow growth and clinical similarity to benign conditions. However, certain subtypes of penile sarcoma such as epithelioid sarcoma are well known for their propensity for local recurrence with regional and distant metastases, making early and correct diagnosis crucial. We report a case of undifferentiated pleomorphic sarcoma of the penis and review the literature.

CASE REPORT

A 59-year-old male initially presented to an outside urology clinic with a pea-sized mass at the base of his penis. The patient denied any hematuria or discharge. He had a history of herpes simplex infection but no history of urinary tract infection or recent travel. The patient denied any constitutional symptoms such as fever, unintentional weight loss, loss of appetite, chills, or night sweats. During physical examination, the mass was free of erythema or ulceration without inguinal lymphadenopathy. Because the lesion appeared to be benign, careful monitoring was recommended. However, the patient presented 6 months later with an unexpectedly enlarged mass. The mass was resected the following month, and the pathology showed high-grade undifferentiated spindle cell sarcoma. Two months later, a restaging positron emission tomography (PET) scan revealed a 1.5-cm recurrence involving the corpora cavernosa. The patient was referred to our department for reevaluation and discussion of treatment options.

After a multidisciplinary conference and discussion with internationally recognized sarcoma experts, the decision was made to treat the patient with chemotherapy followed by restaging and partial penectomy. A chemotherapy trial with 2 cycles of doxorubicin (Adriamycin, 75 mg/m²) and ifosfamide (2,000 mg/m²) was initiated. Before the second cycle of chemotherapy, 2 hypermetabolic suprapubic/peripenile lymph nodes were identified from PET-computed tomography (CT), suggestive of local metastatic disease (Figure 1). After the second cycle of chemotherapy, magnetic resonance imaging (MRI) with and without contrast showed a reniform 1.8 × 1.3-cm (anteroposterior [AP] and transverse [TR], respectively) infrapubic mass near the corpus cavernosum, corresponding to the largest area of hypermetabolic activity on the previous PET-CT (Figure 2). A second smaller lesion was noted on the left infrapubic region, measuring 0.5 × 1.2 cm (AP and TR,

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respectively). The patient underwent excision of the right infrapubic mass and subsequent repair of corporeal defects. Pathology revealed multiple areas of undifferentiated pleomorphic sarcoma in a background of scar tissue (Figure 3).

After the operation, the patient received radiation therapy (66 Gy in 33 fractions) because of his poor tolerance of and lack of response to the previous chemotherapy. After radiation treatment, the patient developed symptoms of cough and shortness of breath. Restaging CT scans revealed multiple bilateral pulmonary nodules (Figure 4), and lung biopsy was positive for undifferentiated pleomorphic sarcoma. Palliative chemotherapy of gemcitabine 900 mg/m² and docetaxel 75 mg/m² was initiated. The patient’s disease rapidly progressed despite treatments, and he died of complications from the metastatic disease 4 months after the resection of his right infrapubic mass.

DISCUSSION
Primary sarcoma of the penis is extremely uncommon, consisting of <5% of all penile cancers. Undifferentiated pleomorphic sarcoma is extremely rare, with only few cases reported in the English literature.

Current treatment options for primary penile sarcoma include tumor excision or partial or complete penectomy for localized disease, as well as radiation therapy and chemotherapy that can be used in combination with surgery in the adjuvant setting or to treat advanced disease. The lack of guidelines and data on the treatment of penile sarcoma complicates the decision-making process during initial therapy. Radiation or chemotherapy alone is considered insufficient for localized disease because of unpredictable variations in patients’ responsiveness to these therapies.
and the need for definitive surgical resection. The benefit of adjuvant chemotherapy in nonmetastatic sarcoma is not clear, requiring additional studies with long-term follow-up, but chemotherapy can certainly be considered in high-risk patients. Our patient exhibited poor response to doxorubicin and ifosfamide as well as to docetaxel and gemcitabine.

Treatment of primary penile sarcoma often occurs late in the disease process because of delays in presentation or diagnosis. Because of the insidious onset of the disease, a patient may not seek medical care for several months to years, waiting until the mass eventually grows and becomes symptomatic. Furthermore, primary penile sarcoma is often misdiagnosed as Peyronie disease or another benign disorder that mimics the symptoms. Some patients present with metastatic disease at the time of presentation, an incurable condition.

The role of systemic therapy for metastatic penile sarcoma is considered similar to that for metastatic sarcomas originating elsewhere. Anthracycline-based chemotherapy regimens such as doxorubicin and ifosfamide or doxorubicin and decarbazine form the basis of first-line therapies, while taxane-based regimens can be attempted in the second-line setting. Newer drugs found useful in treating other sarcomas such as pazopanib, eribulin, and trabectedin may also be considered. Emerging therapies for sarcoma, such as targeted immunotherapies aimed at the programmed cell death protein (PD-1)/programmed death ligands (PD-L1/2) pathway, may also prove useful to treat some patients with primary penile sarcoma.

The best approach for the management of these malignant penile lesions is early diagnosis and treatment before invasion occurs. Widespread patient education regarding the need to seek medical care for new or unusual physical findings or symptoms is needed, and patients’ mindfulness of their own bodies is crucial. Currently, no consensus exists on which patients presenting with penile nodules require further investigation with biopsy. MRI, the preferred method of imaging for delineating soft-tissue tumors, has been found to be unreliable in differentiating between malignant and benign lesions. Although imaging parameters such as patterns of enhancement and tumor necrosis have been considered specific findings that suggest malignancy, studies have shown that these findings may also appear commonly in benign lesions. Because of the rarity of the disease, one may argue against judicious biopsy of all patients presenting with penile nodules. Nevertheless, close and frequent monitoring may benefit patients.

CONCLUSION

Primary sarcoma of the penis is a rare disease, and limited data are available to guide treatment. Resection with or without adjuvant therapies is the standard treatment for the localized disease, whereas patients with advanced disease may be treated with radiation or systemic therapies. However, surgery is considered the only curative treatment, and, therefore, early recognition and surgical excision may improve the outcomes of patients with this disease.

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REFERENCES


Figure 4. Axial computed tomography scan shows pulmonary nodules consistent with metastases (arrow).


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