Pleomorphic malignant fibrous histiocytoma/undifferentiated pleomorphic sarcoma of the glans penis

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Abstract

Primary sarcoma of the penis has an extremely low incidence, and its diagnosis and treatment are unclear. A 55-year-old man presented with an oval-shaped mass of the glans penis, which was treated by a wide excision. The pathologic result revealed an undifferentiated high-grade sarcoma, which was diagnosed as a pleomorphic malignant fibrous histiocytoma/undifferentiated pleomorphic sarcoma based on the World Health Organization classification. There was no recurrence at the12-months postoperative follow-up.

Introduction

Primary tumour of the penis is uncommon. Soft tissue tumours of the penis are rare, with most reported as isolated case reports. The most common benign soft tissue tumour is vascular neoplasm, and the most frequent malignant tumours are Kaposi sarcoma and leiomyosarcoma.[1] Among the malignant soft tissue tumours, undifferentiated high-grade sarcoma of the penis is difficult to find, and the diagnosis and treatment are controversial. We report a case of pleomorphic malignant fibrous histiocytoma/undifferentiated pleomorphic sarcoma of the glans penis, which was treated by a wide excision.

Case report

A 55-year-old man (174 cm, 72 kg, body mass index 23.78 kg/m^2) presented with a 3-month history of a 2.5 × 2.0-cm, painless penile mass. Although he had used an antibiotic ointment for the mass, the size had increased, and the colour had changed from whitish to reddish after frequent manipulation. He had no significant surgical and medical histories. He was a heavy smoker, with a history of more than 2 packs per day for 30 years. Physical examination demonstrated a tenderness and discharge of the mass of the glans penis (Fig. 1). There was no enlarged lymph node on the both inguinal areas. Laboratory examination of blood samples showed a white blood cell count of 11.55 × 10^3/mL (51.8% neutrophils, 1.7% eosinophils). Urinalysis revealed 1 to 4 red blood cells and 0 to 1 white blood cells on high power field. Blood tests for syphilis and human immunodeficiency virus were negative. Abdominal and pelvic computed tomography (CT) scans did not detect any enlarged lymph nodes.

A core biopsy of the mass revealed a sarcoma. To treat and obtain a finial diagnosis, we performed a wide excision of the tumour, including penile skin. Pathologic examination revealed a 6.5 × 2.1-cm ovoid specimen weighing 11 g. The tumour with a well-circumscribed margin showed a round shape with a diameter of 2.6 cm and a depth of 1.2 cm. There was no infiltration into the corpora and urethra. Microscopically, the tumour cells had marked cytological and nuclear pleomorphism, admixed with spindle cells and histiocyte-like cells. Cellularity was high and cellular atypia, nuclear pleomorphism, abnormal mitoses, and areas of tumour necrosis were observed. Immunohistochemically, pan-cytokeratin, HMB-45, LCA, CD30, S-100 protein, smooth muscle actin, CD31, and CD34 were not expressed. Only vimentin and CD68 showed immune-reactivity (Fig. 3). Undifferentiated high-grade sarcoma, with French Federation Nationale des Centres de Lutte Contrele Cancer (FNCLCC) grade 3 (total score 7), was a diagnosis of exclusion.[2] After all other potential mimics had been ruled out by immunohistochemical staining, the final diagnosis was a pleomorphic malignant fibrous histiocytoma/undifferentiated pleomorphic sarcoma based on the World Health Organization (WHO) classification of soft tissue tumours.[3]

During 12 months of follow-up after the operation, the patient showed no evidence of recurrence or metastases on physical examinations and imaging studies including CT scan, which was useful to detect local recurrence and lymph node enlargement.

Discussion

Although soft tissue sarcomas may occur anywhere with an annual incidence of 30 per 1 000 000, primary penile sarcoma is very uncommon.[4] Penile soft tissue tumours comprise 5% of penile tumours, and the incidence of penile sarcoma is 0.6 to 1 out of 100 000 patients in developed countries.[1,5] Penile sarcomas are often slow-growing with a benign appearance, similar to chronic granulomatous inflammation or Peyronie's disease. These sarcomas develop over a wide age range.[6] Among these tumours, Kaposi sarcoma is commonly detected at the glans and prepuce, leiomyosarcoma at the shaft and base of the penis, and rhabdomyoscarcoma at the penopubic junction. The presenting signs and symptoms are subcutaneous mass, penile pain and enlargement, priapism, and voiding difficulty.[1]

The histological type of sarcomas does not always provide sufficient information for predicting the clinical course and for planning therapy. Grading based on histological parameters only evaluates the degree of malignancy and mainly the probability of distant metastasis. The 2 most widely used systems are the United States National Cancer Institute (NCI) system and the FNCLCC system.[7] The FNCLCC system is based on a score obtained by evaluating 3 parameters selected after multivariate analysis of several histological features: tumour differentiation, mitotic rate, and amount of tumour necrosis. Staging of soft tissue sarcomas relies on both histological and clinical information. The major staging system was developed by the International Union against Cancer and the American Joint Committee on Cancer, and appears to be clinically useful and of prognostic value.

Penile sarcomas have been classified as superficial and deep, and proper classification is important for deciding therapeutic methods and prognosis. Although a standard therapy remains elusive because of the extremely small number of cases, the Armed Forces Institute of Pathology recommends that small (<2 cm) lesions be managed with local excision, whereas deeper-seated tumours often require partial or total amputation.[8] Although metastasis is rare, it has a tendency to local recurrence, and it may lethal within a few months.

Fibrohistiocytic tumours of the soft tissue are classified as benign, intermediate, and malignant. Malignant fibrous histiocytoma (MFH) is synonymous with high-grade undifferentiated pleomorphic sarcoma and essentially represents a diagnosis of exclusion.

The current WHO classification recognizes the undifferentiated unclassifiable category of pleomorphic sarcomas, and subdivides it into pleomorphic MFH, giant cell MFH, and inflammatory MFH. These sarcomas constitute about 5% of all soft tissue sarcomas in adults. It occurs more commonly in the extremities, and cases of primary undifferentiated pleomorphic sarcoma of the penis are very difficult to find.[9] El Hayek and colleagues[10] suggest that one possible explanation could be the occurrence of de-differentiation. On the other hand, the tumour could have been pleomorphic from the start.

We present a very rare case of penile sarcoma, which was located at the glans. Hematoxylin and eosin staining of the tumor showed malignant characteristics. We performed several immunohistochemical stains. However, we could not find the exact origin of the tumour. Interestingly, vimentin showed immune-reactivity, which was indicative of an undifferentiated high-grade sarcoma. CD68 reactivity was evident, indicating tumor histiocytes. Finally, our diagnosis was a pleomorphic MFH/undifferentiated pleomorphic sarcoma based on the WHO classification of soft tissue tumors. Although it had a malignant potential with high grade, we did not perform chemotherapy because of the superficial nature of the tumor and the absence of metastasis. After 12 months of follow-up, the patient showed no evidence of recurrence or metastases.

Fig. 1. A 2.5 × 2.0-cm, reddish, oval mass of the glans penis. Thick and folded skin is detected at the distal penis.

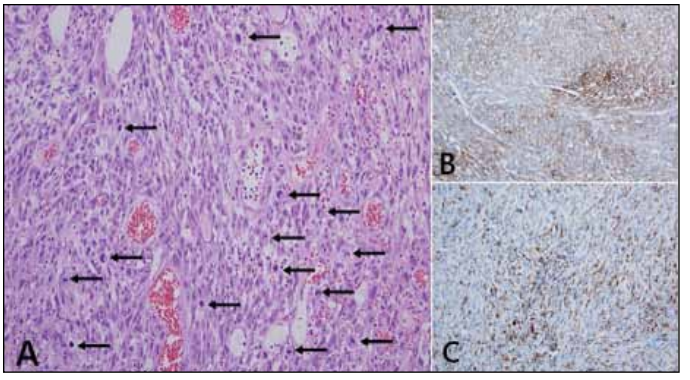

Fig. 2. The tumour is composed of an admixture of spindled and pleomorphic cells with numerous mitotic figures (arrows) (A) and these tumour cells show immunoreactivity to vimentin (B) and CD68 (C).

Conclusion

We report a rare case of pleomorphic MFH/undifferentiated pleomorphic sarcoma of the glans penis. It showed a good prognosis due to an early detection and a wide, local excision for the superficial type. However, it is difficult to determine the diagnosis and treatment because of its rarity.

Competing interests: Dr. Seo, Dr. Oh, Dr. Chuluun and Dr. Choi all declare no competing financial or personal interests.

This paper has been peer-reviewed.

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