Case Report-I

An Unusual Penile Spindle Cell Malignancy

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ABSTRACT

Primary non-epithelial malignancies of the penis are rare. We report a patient with an unusual presentation of an unexpected spindle cell malignancy at this site. A 35year-old man presented with a mass in the shaft of the penis with inguinal lymphadenopathy. A malignant spindle cell neoplasm was seen involving the corpora, consistent with leiomyosarcoma. Immunohistochemically, the tumour was positive for S100, HMB-45 and negative for SMA. A final diagnosis of amelanotic spindle cell melanoma was made. This case is being presented to reiterate the importance of including melanoma in the differential diagnosis of spindle cell sarcomas at uncommon sites.

CASE: A 35-year-old man presented with a progressively enlarging, hard, nodular, ulcerated mass in the proximal shaft of the penis, first noticed 7 months back. He also had left inguinal lymphadenopathy. A biopsy of the lesion done elsewhere was reported as Non-keratinising squamous cell carcinoma, but the slides were not available for review.

His routine investigations were within normal limits. CT scan of the abdomen and penis showed bilateral external iliac and inguinal lymphnodes with right hydrocele and a heterogeneously enhancing penile mass. He underwent a total Penectomy. Gross examination of specimen revealed a ulcerated nodule measuring 5×4 cm in the shaft, the cut surface of which was firm, lobulated and grey-white. The tumour was seen involving the corpora and extending upto glans with free urethral and corporal surgical margins.

On microscopic examination, a malignant spindle cell neoplasm was seen involving the corpora. The tumour cells were arranged in whorls and fascicles, with blunt ovoid nuclei and inconspicuous nucleoli, reminiscent of smooth muscle differentiation. (Fig 1) The mitotic rate

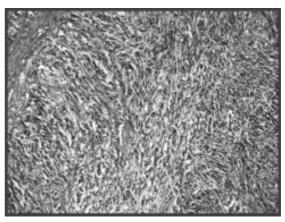


Fig 1. Spindle shaped tumour cells with blunt, ovoid nuclei (H&E x 20)

was 8-9 per 10 hpf and necrosis was noted. The overlying skin was ulcerated. The urethra and corporal margins were uninvolved. A morphological diagnosis of leiomyosarcoma was

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made and immunohistochemistry was advised for confirmation. The tumour was positive for Vimentin, S-100 and HMB-45 and negative for Smooth Muscle Actin and Cytokeratin, ruling out Leiomyosarcoma and spindle cell carcinoma. With the above immunoprofile, a final diagnosis of amelanotic spindle cell melanoma was made. Slides of the main tumour were reviewed. There was no melanocytic proliferation in the overlying skin. FNAC of the inguinal lymph nodes showed metastatic spindle cell neoplasm with a similar morphology. A lymph node dissection was scheduled, but the patient refused any further intervention. A detailed clinical examination did not reveal evidence of a melanoma elsewhere in the body. The patient decided to take further treatment in his hometown.

DISCUSSION

Primary non-epithelial malignancies of the penis are rare.^{1,2} The most common ones have a vascular origin. Kaposi sarcoma and leiomyosarcoma are the commonest mesenchymal malignancies encountered at this site. Leiomyosarcomas are usually seen in mid or late adult life and are more common in the shaft and base of penis. Microscopically, these tumours contain spindled cells with nuclear atypia, mitotic activity and a fascicular growth pattern. On immunohistochemistry, a positive reaction for Smooth Muscle Actin and desmin is noted. Leiomyosarcomas have a greater propensity for local recurrence.

Primary melanomas constitute <2% of all primary penile tumours. Only about 100 cases have been reported in literature, since 1859, mostly case reports.³ The largest series is of 19 cases seen over 25 years.⁴ They are usually seen in ages between 50 and 70 years. 60 – 80% melanomas arise on the glans penis, <10% involve the prepuce and the remainder arises on the skin of the shaft². Most of the primary penile melanomas are pigmented.¹ The common histologic subtypes reported include nodular, superficial spreading and mucosal lentiginous, all of them containing cytoplasmic melanin. The peculiarities of the present case are: a) The

overlying skin showed no melanocytic proliferation b) the tumour was amelanotic and c) Prominent nucleoli, said to be another hallmark of melanoma cells, was absent.

Among prognostic factors, the Breslow index (level of invasion) is the most important, specially tumours of depth above 3.5 mm.^{2,4} Melanomas metastasize early via lymphatic vessels to superficial and deep inguinal lymphnodes and external iliac lymphnodes. As anastomoses are common, there can be bilateral or contralateral involvement¹. Patients with nodal metastases generally die within 2 years.⁴

The treatment of penile melanomas is surgical. When the lesion involves the glans, the suggested treatment is total amputation with bilateral radical groin dissection with or without dissection of the iliac and obturator lymphnodes. The benefit conferred by radiotherapy and chemotherapy in penile melanomas is limited. The overall prognosis in cases of penile melanomas is poor – a 30% 5 year survival. Additional adverse prognostic factors include ulceration and tumour diameter greater than 15 mm.

The correct diagnosis in this case could be established only with the help of immunohistochemistry, as histology was atypical. An awareness of this rare entity and judicious use of ancillary tests are mandated for timely diagnosis.

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