

Primary Malignant Melanoma of the Penis - A Case Report with Review of the Literature.

SV Kumar¹, K Janardhana Rao², KV Ram Prasad³, and Bhagya Lakshmi Atla^{4*}.

¹Department of Surgery, Andhra Medical College / King George Hospital, Visakhapatnam - 530002, Andhra Pradesh, India.

²Department of Surgery, Andhra Medical College, Visakhapatnam - 530002, Andhra Pradesh, India.

³Department of Anatomy, Andhra Medical College, Visakhapatnam - 530002, Andhra Pradesh, India.

⁴Department of Pathology / King George Hospital, Andhra Medical College, Visakhapatnam - 530002, Andhra Pradesh, India.

Case Report

Received: 03/02/2014

Revised: 26/02/2014

Accepted: 07/03/2014

*For Correspondence

Department of Pathology / King George Hospital, Andhra Medical College, Visakhapatnam, Andhra Pradesh, India.

Keywords: Malignant melanoma of penis, penectomy, lymph node metastasis.

ABSTRACT

Primary malignant melanoma of the penis is a rare condition, and about 200 cases of melanoma have been reported in the literature till today since it was first reported in 1859. Here we report a case of malignant melanoma of the penis in a 50 year old male who presented with a pigmented ulcero-proliferative growth on the glans penis with bilateral massive ilio-inguinal lymphadenopathy. The patient underwent fine needle aspiration of the ilio-inguinal lymphnodes and an incisional biopsy of the penile lesion which was suggestive of malignant melanoma with metastatic deposits in the ilio-inguinal lymphnodes. A case of malignant melanoma of penis is presented due to the rarity of the occurrence of malignant melanoma at this site and its dismal prognosis, and to emphasize the importance of its early diagnosis.

INTRODUCTION

The penis is a rare primary site for malignant melanoma. About 200 cases of melanoma of the penis have been reported in the literature since the first case was reported in 1859 [5]. Less than 1% of all primary penile cancers were malignant melanomas. The diagnosis of malignant melanoma of penis is often delayed by the patient's reluctance to consult a physician. The median age of the patients is 65 years with 5 year survival rate of 10%. Early metastatic spread, advanced age at onset and late diagnosis account for the poor prognosis [7,11]. The low number of reported cases does not allow for development of a standardized therapeutic protocol, which results in treating malignant melanoma very often similarly as other malignant tumors of the penis.

Case Report

A 50 year old uncircumcised male patient presented with a brownish pigmented growth on the glans penis of 8 months duration, which is rapidly enlarging in size. On clinical examination a 6 x 5cm ulcero- proliferative growth is noted involving glans penis and bilateral massive ilio- inguinal lymphadenopathy noted (Fig; 1).

Incisional biopsy was done for the penile lesion and it is suggestive of malignant melanoma, FNAC from lymph nodes shows diffusely scattered spindle cells with marked nuclear pleomorphism with brown to black pigment in the cytoplasm, suggestive of melanoma (Figure:2&3). Metastatic survey with computed tomography of chest, abdomen and pelvis were negative. Patient underwent total penectomy with bilateral ilio- inguinal block dissection. On Gross examination the penectomy specimen was measuring 7x4x3 cms ; cut section showed a brownish black ulcero-proliferative growth involving the entire glans penis infiltrating the corpora cavernosa and corpus spongiosum and just abutting the urethral meatus. Gross examination of the bilateral ilio-inguinal lymph nodes showed matted grey brown masses with diffuse brownish black discoloration on cut section. Under light microscopy with Haematoxylin & Eosin (H&E) stained sections showed tumour composed of pleomorphic tumour cells with hyperchromatic nuclei with prominent nucleoli arranged in sheets and fascicles with intra and extracellular brownish black melanin pigment (Fig:4) and sections from all the lymph nodes showed tumour deposits. On histopathological examination the final diagnosis of malignant melanoma of the penis with bilateral

ilio-inguinal lymph node metastasis was made. The patient was followed up for 6 months which was uneventful and was subsequently lost to follow up.



Figure 1: Clinical picture showing a 6 x 5cm ulcero- proliferative growth involving glans penis and bilateral massivelio- inguinal lymphadenopathy.

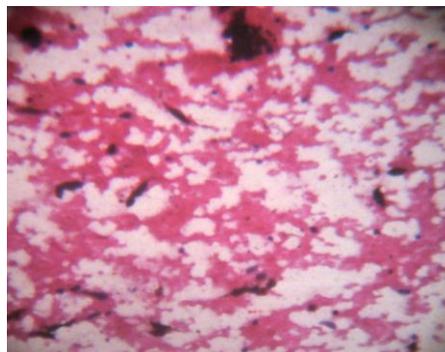


Figure 2: Photomicrograph showing diffusely scattered spindle cells with brown to black pigment granules on cytology. (H&E; 100X)

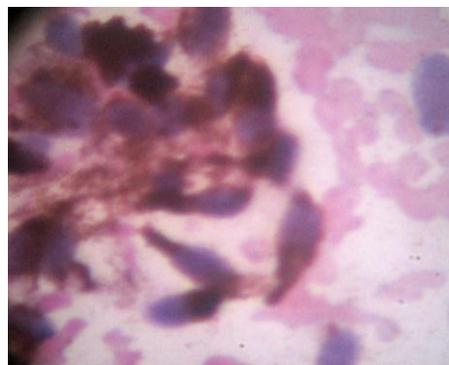


Figure 3: Photomicrograph showing spindle cells with pleomorphic nuclei and brown to black pigment granules both intracellularly and extracellularly on cytology (H&E; 400X)

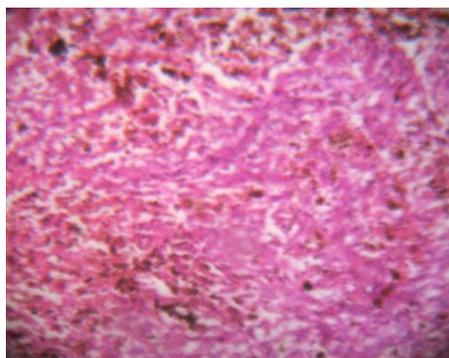


Figure 4: Photomicrograph showing pleomorphic tumour cells arranged in sheets and fascicles with intra and extracellular brownish black melanin pigment. (H&E; 100X)

DISCUSSION

The Dartos fascia and the fascia of the External oblique muscle (Bucks fascia) constitute the superficial and deep fascia of the penis respectively. The body of the penis consists of three erectile bodies – Two corpora cavernosa and the corpus spongiosum. The corpora cavernosa are covered by the tunica albuginea, a dense fibrous tissue is divided by the septum penis. The corpus spongiosum surrounds the urethra and distally becomes the glans penis.

The inguinal nodes draining the penis are divided into two groups: Superficial: 8 to 25, located beneath the subcutaneous fascia which drain the skin of the penis and prepuce, especially into the superomedial zone. The superficial nodes are divided into four quarters by two perpendicular lines drawn over the sapheno femoral junction. They are: superomedial, superolateral, inferomedial, and inferolateral. Deep: 3 to 5 located around the fossa ovalis which drain the lymphatics of the glans and corporeal bodies of the penis.

The Sentinel node is present in the superio-medial zone between the superficial inferior epigastric vein and the superficial external pudendal vein. It is the first lymph node to contain metastatic cancer, anatomically, clinically, and pathologically. The deep node of Cloquet, is the most constant and usually the largest node, found medial to the femoral vein and just underneath the inguinal ligament. The superficial and deep nodes ultimately drain into the pelvic nodes. A communication exists between the left and right lymphatic vessels.

Malignant melanoma of the penis accounts for less than 1% of all primary penile malignant lesions, and less than 0.2% of all extra-cutaneous malignant melanomas in men.^[10] However, it is believed to be even less frequent in circumcised men. It results from malignant transformation of nevus or de novo. It is localized at the glans penis (82%), followed by the prepuce (< 10%), the urethra, meatus, and penile shaft^[1,9]. In contrast to penile squamous cell carcinoma, malignant melanoma of penis is rare in blacks, and it usually affects white men in their fifth and sixth decades of life^[1]. Delayed diagnosis may be due to several reasons such as patient reluctance to seek medical treatment due to denial or fear of the possible malignant change or lesions might be misdiagnosed as an inflammatory process and be treated with antibiotics or various topical ointments or a patient may not have noticed possible malignant changes of a long-existing nevus^[1].

Melanoma presents as a blue black or reddish brown pigmented papule, plaque, or ulceration on the glans penis. It occurs on the prepuce less frequently. Diagnosis is made by histological examination of biopsy specimens, which demonstrate atypical junctional cell activity with displacement of pigmented cells into the dermis and IHC studies for S100, Melanin –A, and HMB-45. The clinical differential diagnosis includes junctional melanocytic naevus, penile melanosis, penile lentigo and atypical pigmented penile macules.

Hematogenous metastasis occur by means of the vascular structures of the corporeal bodies to the liver, lung, bone, brain, and other distant sites; lymphatic spread to the regional inguinal and pelvic nodes occurs by lymphatic permeation. Distant metastatic spread has been found in 60% of patients at the time of initial presentation, which is similar to the case for squamous cell carcinoma of the penis^[6]. Lack of subcutaneous fatty tissue in penile shaft and rich lymphatic vessels relate to the susceptibility to metastasis resulting in poor prognosis.

Clinical staging in most published cases follows that of Bracken and Diokno: Stage I- melanoma confined to the penis, stage II- spread to the inguinal lymph nodes and stage III- distant metastasis^[8]. This staging system, however, ignores 2 other critical prognostic factors: 1) The thickness of the primary lesion is correlated with rates of metastasis and mortality. In lesions less than 0.76 mm thick, the metastatic rate is low (1%) and there is a more-favourable prognosis than with thicker lesions (>1.5 mm) which produce a high risk of vascular and lymphatic invasion & 2) The different levels of invasion of the primary lesion is important, since the thinness of the penile skin may facilitate early dissemination. Stage II and III diseases carry a poor prognosis, mainly because of the lack of effective adjuvant systemic therapy.

Surgery is the primary mode of treatment; radiotherapy and chemotherapy are of only adjunctive or palliative benefit. Treatment modality is determined by tumor stage, patient age and his sexual activity. For stage I melanoma and stage II melanoma adequate excision of the primary tumor by partial or total penile amputation together with en bloc bilateral inguinal and/or ilio-inguinal node dissection has been advocated. For stage I lesions, particularly those with a minimally invasive tumor (< 1.0 mm in thickness) prophylactic regional lymph node dissection is not necessary^[2]. If the primary lesion is greater than 1.5 mm in thickness, prophylactic bilateral superficial lymph node dissection has been recommended as an adjunct to a penectomy even though there is no palpable adenopathy, because about 50% of patients have lymph node metastasis at the time of diagnosis^[11]. For patients with stage II disease, regional lymph node dissection is indicated because of the high incidence of coexisting occult distant metastasis. Furthermore, as compared with squamous cell carcinoma, regional nodes are less effective as barriers to dissemination; hence the prognosis of malignant melanoma penis is worse than for similarly staged squamous cell carcinomas^[11]. Stage III disease carries an extremely poor prognosis. A partial or

total penectomy is done for control of the local complications like bleeding, infection, and urethral obstruction. A regional lymph node dissection is recommended only for palliation, for massive lymph nodes (i.e. pain, mass effect, movement impairment, skin breakdown and tumor fungation; and prevention of erosion of femoral vessels.) [11]. For tumors of the foreskin, circumcision may be adequate [4].

Although sentinel lymph node biopsy techniques are increasingly utilized in more common sites of melanoma, its use in penile melanoma is unproven, and a learning curve of over 25 cases with this rare malignancy may be a barrier [4].

Stage II and III diseases carry a poor prognosis, mainly because of the lack of effective adjuvant systemic therapy [3]. Einhorn et al. reported that patients with disseminated melanoma treated with chemotherapy had a median survival of about 5 months [3].

Combination chemotherapy with (Nimustine Hydrochloride) ACNU, (vincristine) VCR and DTIC (Dacarbazine) showed a response rate of 23% in the patients with disseminated malignant melanoma. Combination chemotherapy is recommended as adjuvant therapy in case of tumor thickness is more than 1.5 mm. Immunotherapy like interferon alfa 2 can be added to chemotherapy. Malignant melanoma penis is relatively radio resistant although radiation therapy has been advocated as a palliation.

The prognosis for patients with penile melanoma is clearly dependent on stage of the primary tumor and the presence or absence of inguinal metastases. Few reports focused on the concept of mucosal site penile melanomas—glans, meatus, fossa navicularis, and distal urethra [7]. These lesions may appear more aggressive than cutaneous lesions, but delay in diagnosis may be a factor. In a pooled, retrospective analysis of 66 cases, the recurrence outcomes were similar for cutaneous melanomas of comparable tumor thickness [7].

CONCLUSION

A rare case of melanoma of penis with massive bilateral ilio- inguinal lymphadenopathy is presented here. The review of literature showed a uniform dismal prognosis.

REFERENCES

1. Begun FP, Grossman HR, Diokno AC, Sogani PC. Malignant melanoma of the penis male urethra. *J Urol.* 1984;132:123-5.
2. De Vita VT Jr, Hellman S, Rosenberg SA. *Cancer: principles and practice of oncology*, 9th ed. Philadelphia: JB Lippincott, 2005 Vol I. Page No 1745.
3. Einhorn LH, Burgess MA, Vallejos C et al. Prognostic correlations and response to treatment in advanced metastatic melanoma. *Cancer Res.* 1974;34:1997-2004.
4. Gerald H. Jordan, Kurt A. McCammon. *Surgery of the Penis and Urethra*. In, Alan J. Wein (ed). *Campbell-Walsh Urology*, 10th edition. Philadelphia, Elsevier publishers, 2012;Vol I. page no 929.
5. Gross SD: *A System of Surgery*, 6th ed. Philadelphia, WB Saunders, 1882.
6. Johnson DE, Ayale AG. Primary melanoma of penis. *Urology.* 1973;2:174.
7. Larsson KBM, Shaw HM, Thompson JF, et al. Primary mucosal and glans penis melanomas: The Sydney Melanoma Unit Experience. *Aust N Z J Surg.* 1999;69:121-6.
8. Muss HB, Richards F, Barnes P et al. Radionuclide scanning in patients with advanced malignant melanoma. *Clin Nucl Med.* 1979;4:516-518.
9. Oldbring J, Miduloroiski P. Malignant melanoma of the penis and the male Urethral. *Cancer.* 1987;59: 581-7.
10. Primus G, Soyer HP, Smolle J et al. Early 'invasive' malignant melanoma of glans penis and male urethra. *Eur Urol.* 1990;18:156-9.
11. Stillwell TJ, Zincke H, Gaffey TA, et al. Malignant melanoma of the penis. *J Urol.* 1988;140:72-5.