

# Epithelioid Hemangioendothelioma Of The Penis: A Case Report

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## Citation

N Ghosh, T Jindal, M Kamal, R Sharma, P Gokulkrishnan, S Mukherjee, S Mondal, D Karmakar. *Epithelioid Hemangioendothelioma Of The Penis: A Case Report*. The Internet Journal of Urology. 2012 Volume 9 Number 4.

## Abstract

Epithelioid hemangioendothelioma is a rare vascular tumor of low malignant potential most commonly found in the lung, liver and soft tissues. Penis is a very rare site for it. We report a case of penile epithelioid hemangioendothelioma. Surgery is the standard treatment and close clinical follow up is necessary due to its unpredictable natural history.

## INTRODUCTION

Epithelioid hemangioendothelioma is a rare vascular tumor. It is characterized by vessels lined by epithelioid endothelial cells along with inflammatory cell infiltrate. It can occur in different locations such as soft tissue, lung, liver, and bone. Penis is a very rare site for it. We report a case of penile epithelioid hemangioendothelioma and review the literature.

## CASE REPORT

A 38 years old male presented with a non-healing ulcer over the glans penis for two months. His past history was unremarkable apart from a history of ceremonial circumcision at one year of age. On examination, there was an ulcerated lesion of size about 1 cm with irregular margins over the left side of glans penis close to corona (Figure 1).

## Figure 1

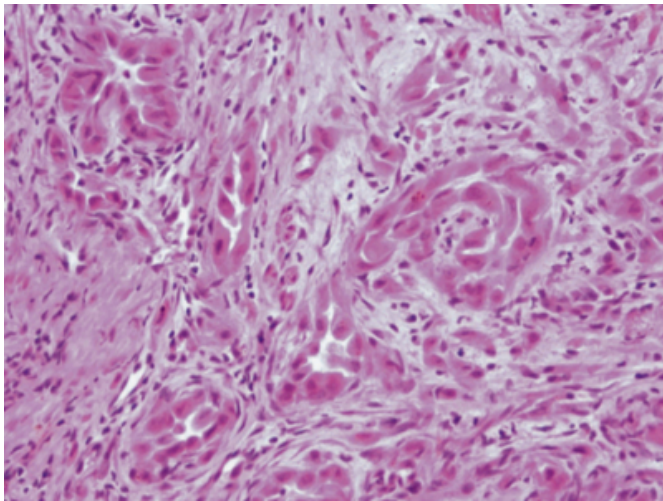
Figure 1: Photograph of the lesion



The base was indurated but there was no tenderness. There was no inguinal lymphadenopathy. Clinically, carcinoma of penis was suspected hence an incisional biopsy was done from the margin of the lesion. Histopathological examination revealed focal ulceration of squamous epithelium with granulation tissue formation (Figure 2).

**Figure 2**

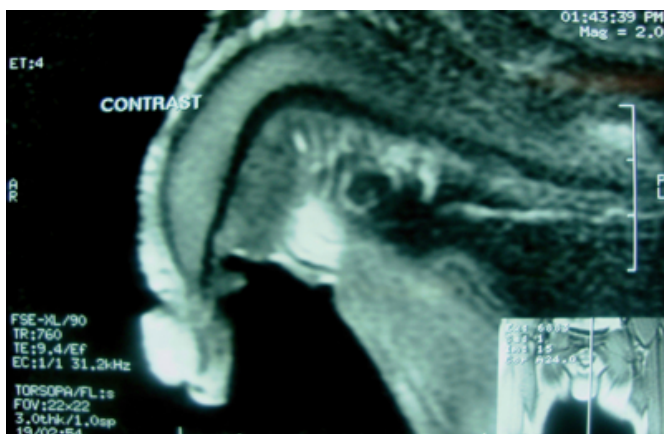
Figure 2: Histopathology picture of the lesion



The sub epithelial tissue was markedly hyalinised and had irregular islands of focally necrotic cells that appeared epithelioid and moderately pleomorphic. Many cells contained intracytoplasmic vacuoles while few contained red blood cells. The cells were embedded in a myxochondroid matrix. There was perivascular and intravascular growth of the tumor cells. The overall appearance was suggestive of epithelioid hemangioendothelioma. Gadolinium enhanced MRI of penis performed at four weeks following biopsy revealed a 2 cm lesion involving the left half of the glans with the major bulk on the dorsal aspect. The lesion appeared brighter in T2 weighted image. There was invasion of the left corpus cavernosa. Right corpus cavernosa and corpus spongiosum were not involved. There was no significant ilio-inguinal lymphadenopathy (Figure 3 and 4).

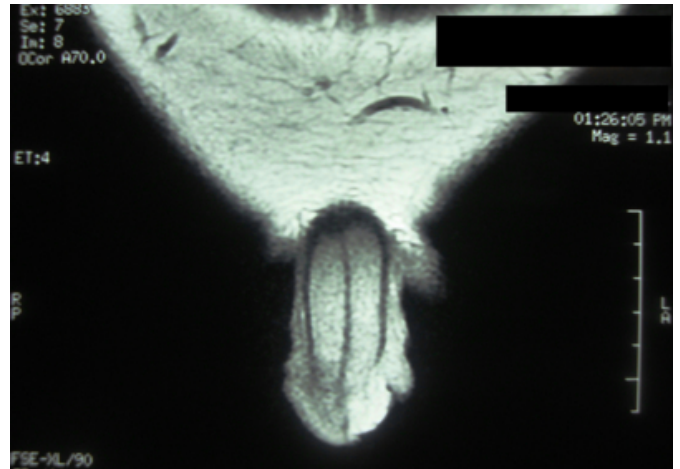
**Figure 3**

Figure 3: Contrast enhanced MRI of the lesion (lateral view)



**Figure 4**

Figure 4: Contrast enhanced MRI of the lesion (cross section view)



Metastatic workup did not detect any distant metastasis. Partial amputation of the penis was planned but the patient refused surgery and was lost to follow up.

## DISCUSSION

Epithelioid hemangioendothelioma is a rare vascular tumor that can occur in different locations such as soft tissue, lung, liver, and bone. Weiss and Enzinger initially described it in 1982 (1). It is a part of vascular proliferations characterized by the presence of epithelioid endothelial cells. It is considered a tumor of intermediate malignant potential between the benign variant called hemangioma and the more aggressive variant called epithelioid angiosarcoma. Epithelioid hemangioendothelioma most commonly involves the soft tissues and occurs as a solitary lesion. It can present with metastases. Rates of metastasis differ according to anatomical locations (20% in soft tissues, 25% in liver and 15% in lungs). Mortality rates also differ according to anatomical locations (13% in soft tissues, 35% in liver and 65% in lungs).

The histopathological differential diagnoses include Kimura's disease and bacillary angiomatosis. Immunohistochemical staining can confirm the endothelial phenotype, neoplastic cells being strongly positive for vimentin, CD31 and factor VIII.

The penis is an unusual location for the disease. There are only eight papers in the English literature that describe the penile involvement. Most penile epithelioid hemangioendothelioma are solitary masses, although multifocal penile epithelioid hemangioendothelioma has been previously described in one report. It usually has an

indolent course. Metastasis is rare.

There is no consensus regarding appropriate imaging for penile epithelioid hemangioendothelioma due to rarity of the cases. One study had used high frequency penile ultrasound to evaluate the lesion (3). Perhaps the first time, we have used Gadolinium contrast MRI to image the lesion – contrast enhanced MRI provided excellent anatomical details of the lesion as well as invasion of corporal bodies; which might be very important in decision making during surgical treatment.

Treatment of localized penile epithelioid hemangioendothelioma without distant metastasis includes wide local excision. Recurrence rates can be as high as 40% (2); hence close follow up with periodic physical examination is warranted. Metastatic epithelioid hemangioendothelioma of penis have been reported (2), but no established management policy due to paucity of cases. Interferon alpha and paclitaxel have been tried with some success (3).

**Figure 5**

Table 1: Different reported series of epithelioid hemangioendothelioma of penis

Series	No of cases	Presentation(s)	Management(s)
Weiss and Enzinger	3	Solitary penile lesions with pain on erection	Local excision
Kamat <i>et al</i>	1	Solitary penile lesion	Surgery, interferon alpha and follow-up of 65 months without recurrence
Wen <i>et al</i>	1	Metastatic three chamber priapism	Paclitaxel, patient died
Gharajeh <i>et al</i>	1	multifocal epithelioid hemangioendothelioma masquerading as superficial penile vein thrombosis	Surgically excised and did not recur after 1 year

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