Case Report

Primary Pseudomyogenic Haemangioendothelioma of The Penis Presenting As A Painful Nodule

Mohammed Shahait¹, Aya Khairalla², Rami Nasr³*

¹Division of Urology, American University of Beirut Medical Center, Beirut, Lebanon
²Department of Pathology, American University of Beirut Medical Center, Beirut, Lebanon
³Division of Urology American University of Beirut Medical Center, Beirut, Lebanon

*Corresponding author: Rami Nasr, Division of Urology, American University of Beirut Medical Center, Beirut, Lebanon; E-mail: ww04@aub.edu.lb


Received Date: 14 December, 2016; Accepted Date: 22 December, 2016; Published Date: 29 December, 2016

Background

Most primary penile tumors are predominantly malignant epithelial tumors, usually squamous carcinoma. Herein, we report on case of isolated penile PMHE treated by penile-preserving surgery.

Case presentation

57-year-old gentleman referred to our clinic with painful penile lesions, mainly involving the glans; after being treated by empirical antibiotic. Physical examination revealed multiple tender nodules over the penis with no apparent skin changes. Punch biopsies were taken from these nodules, and the final pathology was consistent with epithelioid haemangioendothelioma (PMHE). MRI of the penis & FDG-PET scan showed no evidence of metastatic disease. We performed a penile-preserving surgery aiming to achieve a trifecta of negative surgical margin, functional organ, and aesthetically acceptable penis. Intra-operative resection margins were negative. The patient had a smooth postoperative course. At one month follow-up, the patient was able to have a normal sexual activity with complete penetrance. After 15 months from the surgery, follow-up MRI showed no recurrence of the disease.

Conclusion

Penile preserving surgery is a feasible option in treating pseudomyogenic haemangioendothelioma (PMHE) of the penis.

Most primary penile tumors are predominantly malignant epithelial tumors, usually squamous carcinoma. Several uncommon epithelial and soft tissue tumors have been described. Malignant soft tissue tumors are rarely described in the penis; the majority of the sarcoma cases are Kaposi sarcoma in patients with acquired immune deficiency syndrome (AIDS). Nevertheless, another rare form of sarcoma also has been reported such as angiosarcomas, epithelioid haemangioendotheliomas, epithelioid sarcomas, leiomyosarcomas, undifferentiated pleomorphic sarcomas, rhabdomyosarcomas, malignant Schwannomas [1].

Amid the advancement in the histopathology field along with the introduction of Immun histochemistry and gene profiling, this led to the reclassification of soft tissue sarcoma and an introduction of new variants such as pseudomyogenic haemangioendothelioma (PMHE) [2]. PMHE is a rare tumor. It is a soft tissue tumor with unique clinical and pathological features. Usually, it affects the limbs of young adults with male predominance; however, it is on the odd occasion involving the penis. Herein, we report a very rare case of isolated penile PMHE treated by penile-preserving surgery.

CASE

An otherwise healthy 57-year-old gentleman referred to our clinic with painful penile lesions, mainly involving the glans; after being treated by empirical course of antibiotic. Physical examination revealed adult circumcised penis, with no apparent skin changes. Upon palpation, multiple tender nodules over the shaft and the glans were felt. There was no palpable inguinal lymph node, and the digital rectal exam was normal. At that juncture, we elected to perform punch biopsies of the lesions. Pathology revealed nests of atypical epithelioid cells within a fibrous background, all consistent with epithelioid sarcoma. The unusual presentation spurred us to run a special immunohistochemistry staining; which showed that tumor was strongly positive to keratin AE1/AE3, ERG, and INI-1, along with multifocal positivity for CD31, while EMA is negative (Figure 1).
Figure 1: Subcutaneous penile tumor made of cellular nodules merging in a collagenous stroma (A (x4) & B (x10), H&E). Nodules are composed of spindle and polygonal cells with eosinophilic cytoplasm, mild atypical nuclei and prominent nucleoli admixed with neutrophilic infiltrate (C (x40), H&E). The tumor cells are diffusely positive for cytokeratin (D, CKA1-CKAE3), negative for CD34 (E) and positive for SMA (F).

These findings along with Dr. Fletcher input made our pathologist to reconsider their reading and to entertain the PMHE as an alternative diagnosis. MRI of the penis & FDG-PET scan showed no evidence of metastatic disease (Figure 2, 3).

Figure 2: FDG-PET scan demonstrates high uptake by the nodule.

Figure 3: MRI demonstrates no inguinal lymphadenopathy.

Given the low metastatic potential of this tumor, we opted to perform a penile-preserving surgery aiming to achieve a trifecta of negative surgical margin, functional organ, and aesthetically acceptable penis. Wide local excision of the lesions was performed (Figure 4).

Figure 4: Intraoperative picture. A) Glans penis defect after wide excision of multiple nodules. B) Glans after reconstruction.

Intra-operative resection margins were negative. The patient had a smooth post-operative course. At one month follow-up, the patient was able to have a normal sexual activity with complete penetrance.

Outcome And Follow-Up

Twelve months after organ-preserving surgery for his penile PMHE, the patient is still disease free with no evidence of local recurrence of distant metastasis. Patient has been followed up by MRI pelvis which showed no evidence of disease persistence or recurrence. At one year after his surgery, he will be followed up with a second MRI and a PET scan for a complete assessment.

Discussion

PMHE is a rare variant of sarcoma. In 2011; Fletcher et al identified a subgroup of patients with epithelioid sarcoma with distinctive pathological features, explicitly, are abundant myoid-appearing spindle cell in the morphology, expression of FLI1, common reactivity for CD31, lack of epithelial membrane antigen, CD34, and PAN-K expression, and intact INI1. Moreover, the baleful presentation in addition to the benign course of this subgroup of patients made the authors pose a new entity for this variant “Pseudomyogenic hemangioendothelioma” [2]. Thus, in the 2013 WHO classification of soft tissue tumors, the PMHE was recognized as a distinct entity [3].

So far there are 70 cases of PMHE have been described in the literature. Only two cases had penile involvement, in one instance the lesions were exclusively sequestered in the penis [2]. It is a disease of a young adult where it is rarely described above age...
40, intriguingly, with male predominance. Characterized by multicentricity; in addition to that, it can involve the entire soft tissue plane and in rare cases the bone [4].

PMHE has a tendency to recur locally with low potential to metastasize distally. Therefore, a vast preponderance of cases was treated by local excision, even in multifocal scenarios [2, 5]. Unfortunately, there is no consensus on the follow-up duration, as there are reported cases in the literature where the patient developed distant metastasis years after the primary surgery [5].

PMHE is a soft tissue tumor with low metastatic potential. High clinical suspicion along with distinctive pathological features should help in differentiating this tumor from sarcomas, mainly, epithelioid variant. Taking into account, the indolent behavior of this tumor organ-preserving surgery is the mainstay of treatment.

References