A case of primary extraosseous penile osteosarcoma and review of the literature

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ABSTRACT

Primary extraskeletal osteosarcoma of the penis (ESO) is a very rare condition. As per our knowledge, this is the seventh reported case in the world literature. We report a case of primary ESO of the penis in a young male, which was treated with surgery and adjuvant chemotherapy. This case is presented for its rarity and the scarcity in the literature regarding its treatment and prognosis.

Key words: Desmin, primary extraskeletal osteosarcoma of penis, vimentin

INTRODUCTION

Primary osteosarcomas have been reported in a number of extraosseous sites as retroperitoneum, thigh muscles, and in a variety of other sites. But primary ESO in the penis is a rare condition. About six cases have been reported in the world literature which are cited in Table 1.

CASE REPORT

A 35-year male presented with a hard mass at the base of the penis since the past 1 year with rapid growth over the past 4 months. It was painless. He had no voiding difficulty. There was no difficulty in erection or ejaculation. On evaluation, there was a hard mass at the base of the penis on the dorsal and the ventral aspects of the penis reaching up to the pubic symphysis. CT scan of the pelvis showed extensive ossification of the penis [Figure 1]. He underwent a biopsy of the mass, which showed spindle-shaped cells with nuclear anaplasia and pleomorphism and osteoid formation suggestive of osteosarcoma of the penis [Figure 2]. Immunohistochemistry showed desmin, vimentin, and SMA + vity and S-100 and cytokeratin were –ve, which confirmed the diagnosis of osteosarcoma of the penis [Figure 3]. Bone scan and CT scan of the chest were negative for metastases. The patient underwent a total penectomy with urinary diversion [Figure 4]. He underwent three cycles of adjuvant chemotherapy. On follow-up for 1 year, the patient has no local or systemic recurrence.

DISCUSSION

Primary extraskeletal osteosarcoma (ESO) is an exceedingly rare condition, accounting for just 1% of soft-tissue sarcomas. Only six cases of primary ESO of the penis have been
reported in the literature.\cite{1-6} In contrast to the primary osteosarcoma of the bone, ESO of the penis affects middle aged to elderly patients. Theories regarding its etiology include trauma, radiation, and dermatomyositis; there is no evidence to implicate any of these factors in this patient.

Almost all ESO are high grade. Malignant osteoid, bone, and cartilaginous elements are found. Histological subtypes and ultrastructural appearances are same in osseous and extrasosseous tumors; careful histological assessment is necessary and immunohistochemistry may sometimes be helpful. The traditional treatment for ESO is surgery alone or combined with chemotherapy. So this patient was treated with a total penile amputation to obtain adequate resection margins. Osseous osteosarcoma is a relatively radioresistant tumor, so adjuvant radiotherapy was not given for this patient due to its relative radioresistivity and presence of clear surgical margins. The role of adjuvant chemotherapy in ESO is debatable. Cisplatin-based chemotherapy alone or combined with surgery had limited clinical benefit over the traditional treatment.

The overall prognosis for patients presenting with ESO of the penis is poor with a 5-year survival rate of < 25%; 36% develop local and 65% develop distant metastases.

Of the six cases presented, one had areas resembling malignant fibrous histiocytoma\cite{3} and other was a mixed tumor with osteosarcoma and squamous cell carcinoma. Of the cases described in literature, two patients underwent a partial penectomy,\cite{3,5} while two underwent a total penile amputation.\cite{4,6} Bastian et al. described a case of primary ESO of the glans penis, which was managed with a glansectomy with glanular reconstruction.\cite{1}

Only one patient with osteosarcoma has remained alive for

![Figure 2](image2.png)  
**Figure 2:** H & E ×400 showing the presence of spindle-shaped cells with anaplasia and areas of osteoid formation

![Figure 3](image3.png)  
**Figure 3:** Immunohistochemistry showing vimentin positivity

![Figure 4](image4.png)  
**Figure 4:** Total penectomy specimen showing extensive ossification in the corpora cavernosa

| Table 1: The review of literature of the primary penile osteosarcoma cases by various authors |
|---------------------------------|---------|-----------------|-----------------|-----------------|-----------------|
| Ref no. | Authors | Age of patient | Location of lesion | Treatment | Follow-up | Survival |
|---------|---------|----------------|--------------------|-----------|-----------|----------|
| 1.      | Bastian | 46 years       | Glans penis        | Glansectomy with reconstruction | 11 months | No local/systemic recurrence till 11 months follow-up |
| 2.      | Fraser  | 60 years       | Glans penis        | Total penectomy with perineal urethrostomy | 3.5 years | No local/systemic recurrence till 3.5 yrs of follow-up |
| 3.      | Baceti  | 84 years       | Shaft              | Total penectomy with perineal urethrostomy | 1 year | Death due to metastases |
| 4.      | Sacker  | Middle age     | Shaft              | Total penectomy with perineal urethrostomy | 1 year | Death due to metastases |
| 5.      | Edwards | Middle age     | Shaft              | Total penectomy with perineal urethrostomy | 1 year | Death due to metastases |
| 6.      | Shringarpure | 35 years | Shaft | Total penectomy with perineal urethrostomy + chemotherapy | 1 year | No local/systemic recurrence till 1 yr |

Shringarpure, et al.: Primary penile osteosarcoma
3.5 years after surgery. In case of three of other previously reported patients died within 1 year of diagnosis,[3-5] and there is no follow-up information on the fourth.[6] Our patient remains alive and well and free from recurrence 1 year after treatment with surgery and chemotherapy.

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