Rare Malignant Tumors of Clitoris—a Case Report

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Abstract Tumors of the clitoris are very rare. Isolated case reports are available in literature. This case report highlights the importance of this disease, due to its high malignant potential and treatment options. We had come across a rare tumor of the clitoris in a young girl, reported to be malignant extrarenal rhabdoid tumor and patient had a short life span of 6 months.

Keywords Clitoris · Malignant tumors · Rhabdoid tumor · Vulva · Clitoridectomy

Introduction

Tumors of the clitoris are very rare [1, 2] and they are important because of its aggressive nature of clinical course leading to early death in young patients. A rare case of extrarenal rhabdoid tumor of the clitoris is reported here. Similar case was reported earlier, in an elderly patient. These tumors are diagnosed by histology and immunohistochemistry. They need aggressive surgical care with radiotherapy and chemotherapy for better survival [3, 4]. Even with aggressive therapy the prognosis is very poor.

Case Report

A 17-year-old nursing student presented to a physician with painful swelling of the clitoris suspecting infection. Clinical

examination revealed a firm mass arising from the clitoral area and mobile over the bone (Fig. 1). With a clinical diagnosis of fibromatous tumor, the patient was investigated with pelvis X-ray, chest X-ray, and USG of the abdomen, and there was no positive findings. FNAC was done and the smear showed a few spindle cells with large nuclei eccentrically placed suggestive of some aggressive sarcoma. So, patient was investigated and subjected for wide excision.

Operative Findings A firm to hard mass replacing the clitoris with no fixity to the deeper bones or soft tissues was noted. The lesion was arising from the corpora of clitoris and it was widely excised (Figs. 2 and 3).

Gross Findings A firm mass of size 5 cm in diameter surrounded by normal connective tissue was noted. The tumor on division showed a well-defined whitish lesion with uniform appearance (Fig. 4).

Histopathology Findings Sheets of poorly cohesive cells, large polygonal or spindled cells with abundant glassy eosinophilic cytoplasm, paranuclear eosinophilic inclusions, weakly

Fig. 1 Enlarged clitoris with smooth surface

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Fig. 2 Excised mass of clitoris and tumor

PAS positive, keratin positive, and vimentin positive were shown (Fig. 5). Large eccentric vesicular nucleus, round and bean shaped and large, prominent nucleolus, with high rate of mitotic figures were observed. The resected margin was free of tumor infiltration with clearance of 1.5 cm.

Impression The studies are suggestive of malignant extrarenal rhabdoid tumor with resected margin free of tumor.

Even though patient was advised a radical resection with reconstructive procedure, she was not willing for a mutilating surgery. Hence, patient was given radiation and chemotherapy. But 6 months later, the patient expired.

Discussion

Our patient was diagnosed to have malignant extrarenal rhabdoid tumor, one of the rarest tumors of the clitoris reported in literature. There are two similar cases reported in elderly patients [1–3]. Bokhman et al. and Popovici et al. reported few cases of clitoral tumors and highlighted rarity of the clitoral tumors [4, 5].

Fig. 3 Post-excision defect

Fig. 4 Cut section showed whitish appearance

Malignant Extrarenal Rhabdoid Tumor Malignant rhabdoid tumors of the clitoris are neoplasms which show aggressive behavior and a dismal prognosis. A case of malignant rhabdoid tumor of the clitoris occurring in an elderly patient was reported by Haidopoulos et al. [2]. Bond et al. reported a case of alveolar rhabdomyosarcoma of the clitoris in an old patient [2, 3]. Malignant rhabdoid tumor of the clitoris has similarities with other low differentiated tumors; hence, immunohistochemical and ultrastructural assessment should be conducted so that accurate diagnosis is achieved. Individualized extensive surgical treatment might decrease relapsing disease and may improve the prognosis.

In general, malignant rhabdoid tumor is a rare soft tissue neoplasm that occurs in children and young adults. Even though the rhabdoid tumors were initially considered as renal tumors, it can occur in the brain, meninges, ovary, lungs, heart, and external soft tissues. These are highly malignant, aggressive tumors and lethal with median survival of 19 months and 1 year [1–3]. Metastases occur early to lungs, liver, and lymph nodes.

They are having some specific features: (a) a tumor composed of non-cohesive single cells, clusters, or sheets of large tumor cells with abundant glassy eosinophilic cytoplasm, an eccentric vesicular nucleus, and an extremely large nucleolus; (b) positivity for vimentin and/or cytokeratin or other epithelial markers by immunostaining; (c) exclusion of other

Fig. 5 Microscopic view of the tumor (received from pathologist)

tumor types with rhabdoid inclusions (melanoma, other sarcomas, carcinoma). S100 marker will be positive in these cases. (d) They may be weakly PAS positive and keratin positive. Ninety-five percent of rhabdoid tumors will show positive vimentin and 58 % pan-cytokeratin. They are negative for S100.

In malignant extrarenal rhabdoid tumor (MERT), the composite form is characterized by the presence of both a rhabdoid component and other distinctive pathological components, such as transitional cell carcinoma, adenocarcinoma, renal cell carcinoma, various sarcomas, or malignant melanoma but pure form is seen only with rhabdoid tissue.

However, Ota and colleagues suggested that the rhabdoid tumor cells were derived from a primitive pluripotent cell that had the potential for a wide range of differentiation and accounted for the phenotypic heterogeneity observed in MERT [6]. There may be a tumor suppressor gene inactivation or deletion/mutation of a gene at chromosome 22q which might be involved in the genesis of MERT [7–9].

Other Tumors of the Clitoris The histology of a primary carcinoma of the clitoris was found to be different from squamous cell carcinoma of the vulva. It has areas showing a transitional cell carcinoma pattern with foci of spindle and anaplastic cells. The clitoral neoplasm was found to differ in its cytokeratin 3 profile from that described in invasive squamous cell carcinoma of the vulva, especially with regard to the widespread staining for cytokeratin [10, 11].

The commonest tumors of the clitoris are secondaries from the bladder and urethra, which shows features of transitional cell carcinomas with lymph node metastasis [12]. The vulval carcinoma extending to the clitoris is not rare.

Other isolated case reports are epithelioid hemangioendothelioma [13], arteriovenous hemangioma [14], congenital hemangiopericytoma [15], angiokeratoma [16], nevus lipomatosus cutaneous superficialis [17], squamous cell carcinoma, plexiform schwannoma, and Paget's disease of the clitoris.

Treatment of Malignancies of Clitoris

Clitoridectomy combined with radiation and chemotherapy is the choice of treatment [18, 19]. But radical resection is done for rare malignancies of the clitoris. Radical resection of the clitoris and mons with local lymphadenectomy is a very mutilating procedure and the reconstruction requires a bilateral medial thigh flaps or anterolateral thigh flaps as free flaps. Irrespective of the choice of treatment, all the malignancies are having very poor prognosis. Our patient was not willing for the mutilating surgery and hence after wide resection subjected for radiotherapy and chemotherapy.

External beam radiation therapy is often avoided in the treatment, in young children, because of the long-term sequelae. Conventional brachytherapy can reduce these problems, but its use is limited in young children because of radiation exposure to parents and caregivers. Nag et al. [19] reported the first use of high-dose rate remote brachytherapy to treat such cases.

Conclusion

A rare case of malignant extrarenal rhabdoid tumor of the clitoris is reported here. The prognosis of malignant extrarenal rhabdoid tumor is poor even with aggressive surgery chemotherapy and radiotherapy.

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