Botryoid rhabdomyosarcoma of cervix - A Rare case report

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ABSTRACT

Botryoid Rhabdomyosarcoma of the female genital tract is rare in the cervix. It tends to occur in children and young women and has comparatively better prognosis than sarcoma botryoides of the vagina and uterus. We report a case of 20 year old female with Botryoid Rhabdomyosarcoma presenting as a cervical polyp. Only a few cases are reported in literature till now. The definitive diagnosis can be confirmed by histopathology only.

Keywords: Sarcoma botryoides, cervix, polyp.

Introduction:

Rhabdomyosarcoma is an aggressive malignant tumor arising from embryonic mesenchyma and is the commonest soft tissue sarcoma in childhood and young adults. It represents 4-6% of all malignancies [1]. The most common sites of origin for rhabdomyosarcoma are head and neck (35%) followed by the genitourinary tract (25%), extremities, trunk, retro peritoneum and other rare sites (Gastro intestinal tract, perianal and anal). The most common favorable variant is embryonal rhabdomyosarcoma which includes botryoides, spindle and anaplastic histological subtypes. The other two types of rhabdomyosarcoma are alveolar, and pleomorphic variants. These are less common and have less favorable prognosis. The embryonal botryoides variant is associated with a much more favorable outcome than the alveolar and undifferentiated subtypes which are associated with a poor prognosis [2]. Cervical rhabdomyosarcoma is very rare and the largest series in the literature consists of 14 cases only [2-3]. The mean age at diagnosis of patients with cervical lesion is higher than that of patients with vaginal lesions. Primary vaginal lesions are more common than cervical lesions [4]. We report a case of sarcoma botryoides of the cervix presenting as a cervical polyp in 20 year old female.

CASE REPORT

A 20 years old female patient presented with the history of vaginal bleeding of 4 months duration. There are no associate significant symptoms except weight loss. On general physical examination no abnormality was detected. Vaginal examination revealed a mass protruding
through introitus. Biopsy was taken from the mass and sent to pathology department in 10% formalin for histopathologic examination. Macroscopic examination showed multiple grey to white soft tissue bits measuring approximately 1.5 cm in size. Differential diagnosis considered by the gynecologist was rhabdomyoma, edematous cervical mesoderm polyp (Pseudo sarcoma botryoides), adeno sarcoma . On microscopic examination of the lesion, it showed polypoidal lesion lined by stratified squamous epithelium. Underneath the epithelium, band of closely packed tumor cells having hyper chromatic round to oval nuclei with scant cytoplasm were noted (Cambium layer). Tumor cells are concentrated around endocervical glands. Adjacent to this layer hypo cellular areas were noted which showed myxoid degeneration. Congested blood vessels and chronic inflammatory cells were also noted. One focal area showed areas of necrosis and hemorrhages (Fig. 1 and Fig. 2).

Immunohistochemical staining was positive for vimentin and negative for estrogen and progestrogen receptors. Post operative period was uneventful and 2 months follow up of the case showed no recurrence.

**DISCUSSION**

Embryonic rhabdomyosarcoma commonly affects the genito urinary tract, and the vagina is the most common site [5]. However cervix is rate site and accounts for 0.5% only in girls [2]. The most common subtype of embryonic rhabdomyosarcoma is designated as sarcoma botryoides. The exact etiology and pathogenesis of this rare tumor is not clearly known. The majority of rhabdomyosarcoma cases occur sporadically with no specific predisposing risk factors, although a minor proportion of the case may be associated with genetic conditions. Most of them present with a soft grape like clusters, as single or multiple polyps [2]. It is associated with symptoms of leucorrhoea, bleeding and presence of a vaginal mass [2].
The gross examination reveals the polypoidal growth with a grapes like appearance with smooth, glistening surface and areas of hemorrhages [6]. Microscopic examination shows polypoidal lesion lined by stratified squamous epithelium. Beneath intact epithelium condensed sub epithelial layer of rhabdomyoblasts are scattered in a loose myxoid or dense collagen. The cells in cambium layer are round to spindle with oval nuclei with open chromatin and inconspicuous nuclei. The pathological differential diagnosis of this lesion includes rhabdomyoma, edematous cervical mesodermal polyps (pseudo sarcoma botryoides) and adeno sarcoma [7] 

Mesoderm stromal polyp (pseudo sarcoma botryoides) is benign exophytic lesion occurring in the vagina and cervix of women of reproductive age. Histology shows the polypoidal lesion composed of edematous stroma covered by a benign appearing stratified squamous epithelium. The stromal component is composed of bland appearing plump stromal fibroblasts. Mesodermal stromal polyp can be differentiated from sarcoma botryoides by the absence of mitotic figures, lack of rhabdomyoblasts and lack of cambium layer.

Adeno sarcoma arising from uterine cervix may present as a cervical polyp. Histologically the polypoidal cervical lesion shows dense malignant spindle stromal cell proliferation around benign endocervical glands. Stromal cells show marked pleomorphism. Immunohistochemical examination of stromal cells is positive for muscle specific actin, desmin and estrogen receptors. Adeno sarcoma is distinguished from sarcoma botryoid by the leaf like pattern of glands, the fibrous stroma and oestrogen receptor positivity.

Rhabdomyoma is a rare benign tumor of cervix observed in young and middle aged women. Microscopically the lesion shows undifferentiated spindle shape cells with abundant eosinophilic cytoplasm and scattered muscle fibers with in myxoid matrix, beneath intact squamous epithelium. It can be differentiated from embryonal rhabdomyosarcoma by the absence of cambium layer and mitotic figures. Sarcoma botryoides of the cervix behaves less aggressively than sarcoma botryoides of the vagina and uterus[7]. Recently the approach of management of embryonic rhabdomyo sarcoma has been increasingly conservative in order to preserve the fertility in young nulliparous women [3,8].

Intergroup Rhabdomyosarcoma study group staging (clinical classification) system for Rhabdomyo sarcoma is as follows:

**Stage** | **extent of disease, margin status and resectability**
--- | ---
I A | Tumor confined to site of origin and the tumour is resected completely
I B | Tumor infiltrating beyond the origin site but was resected completely
II A | Tumor gross total resection was done but residual microscopic disease still present
II B | Extensive tumor locally (spread to regional lymph nodes present) resected completely
III A  Localized / locally extensive tumor gross residual disease still present

III B  Localized / locally extensive tumor gross after resection major (debulking of >50% of tumor)

IV  Primary tumor of any size with or without regional lymph node involvement with distant metastases irrespective of surgical approach to tumor primary.

Fertility sparing surgery followed by chemotherapy is the appropriate treatment for patients with local disease according to Intergroup Rhabdomyosarcoma study group [9]. Fertility sparing surgery should not be considered with the presence of metastasis (uterine involvement). Surgical treatment (hysterectomy), chemotherapy and radiotherapy used for patients with deep myometrial invasion and lymphatic invasion. The prognosis of sarcoma botryoides of the cervix is better satisfactory than rhabdomyosarcomas of occurring in any other foci of genital system. In the Intergroup Rhabdomyosarcoma study group, patients had a 5 years survival rate of approximately 70% with gross residual disease (stage III) compared with a greater than 90% 5 years survival rate for no residual tumor after surgery in patients (stage I) and approximately 80% 5 years survival rate for patients with tumor gross total resection but microscopic residual disease (stage II)[10]. We report the case due its rarity of presentation in cervix. In our case patient treated with fertility sparing surgery (polypectomy) and diagnosed in early stage of the disease.

CONCLUSION
Embryonal rhabdomyosarcoma of cervix is extremely rare malignancy in young women. Early diagnosis and treatment spares fertility in young women. Early disease stage at diagnosis is a highly favorable prognostic factor.

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