A Rare Presentation of Aggressive Angiomyxoma as a Cervical Polyp

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ABSTRACT

A rare case of aggressive angiomyxoma (AAM) is reported here for its unusual presentation as a cervical polyp, in a 45-year-old woman. It is a soft tissue tumor. Preoperative diagnosis was uterine cervical polyp. Surgical excision was done. Histopathology examination revealed vascular, poorly circumscribed tumor mass composed of spindle cells embedded in myxoid stroma. In the reviewed literature, a single case of AAM mimicking cervical polyp has been reported. Our case may be the second rare case of AAM presenting as a cervical polyp.

KEY WORDS: Aggressive angiomyxoma, angiomyofibroblastoma, cervical polyp, superficial angiomyxoma

INTRODUCTION

Aggressive angiomyxoma (AAM) was first described in 1983 by Steeper and Rosai. This mesenchymal tumor arises from connective tissue of lower pelvis or perineum and has a locally aggressive course. The neoplasm predominantly affects reproductive age females with peak incidence during the third decade of life. The female to male ratio is 6:1. In women, vulvar region is the most common site of involvement.

CASE REPORT

A 45-year-old female, presented to Gynecology Out-patient-Department with complaints of something coming out of vagina since 2 months and yellowish discharge from the vagina since 15 days. Her menstrual cycles were regular. There was no pallor and icterus.

Systemic examination

P/A – Soft, nontender, no organomegaly was present. Per speculum examination showed 6 × 6 cm, polypoid, pedunculated, nontender mass arising from the posterior lip of cervix. Vaginal examination showed normal sized uterus. Hemogram was within normal limits. Patient was negative for human immunodeficiency virus (HIV) and hepatitis B surface antigen (HBsAg). Considering the clinical diagnosis of cervical polyp, excision was performed. Tumor mass was sent to pathology department for histopathological examination.

Gross pathology

On gross examination, the mass was polypoid, grayish-white, and soft to firm, measuring 6.5 × 5.5 × 4 cm [Figure 1]. The cut surface was slimy, gelatinous, and mostly solid with some cystic areas.

Microscopy

Sections revealed poorly circumscribed tumor mass, partly covered by ectocervical stratified squamous epithelium. The tumor mass was composed of spindle and stellate shaped cells with ill-defined cytoplasmic borders, numerous, variable sized thick muscular and thin walled blood vessels, embedded in the abundant myxoid stroma [Figures 2 and 3]. Occasional endocervical gland was also present, in the tumor mass. Cellular atypia was not seen. No cellular pleomorphism, anisonucleosis, increased mitotic activity, or necrosis were seen. No lipoblasts or nerve sheath elements were present. Histological diagnosis of AAM of cervix was given.

DISCUSSION

AAM is a slowly growing myxoid neoplasm that occurs chiefly in the genital, perineal, and pelvic regions of adult women. The neoplasm predominantly affects reproductive
age females with peak incidence during the third decade of life. The female to male ratio is 6:1. In women vulvar region is the most common site of involvement.[3] In the reviewed literature, a rare case of AAM mimicking cervical polyp has been reported by Paplomata et al.[4] Our case may be the second such rare case of AAM presenting as a cervical polyp. Our patient was a 45-year-old female. Size of the tumor was more than 6 cm in diameter. The tumor presented as slowly growing, painless, polypoid, pedunculated mass. Microscopic examination showed poorly circumscribed tumor mass composed of variable sized thick and thin walled blood vessels, abundant myxoid stroma, and uniform bland spindle and stellate cells. Considering all these findings, histopathological diagnosis of aggressive angiomyxoma of cervix was given.

AAM must be distinguished from the more common benign and malignant myxoid tumors including myxoma, myxoid liposarcoma, myxoid neurofibroma, myxoid leiomyoma, leiomyosarcoma, myxoid liposarcoma, myxoid malignant fibrous histiocytoma, and botryoides rhabdomyosarcoma. AAM may also be clinically misdiagnosed as polyps, myxoma, lipoma, and Bartholin’s cyst of vagina. The diagnosis of angiomyxoma may be difficult to establish. The distinctively striking vascular component in AAM helps to rule out the above mentioned neoplasm as differential.[5]

In our tumor, histologically presence of striking vascular component and absence of nests of small basophilic cells (stromal cells) as well as absence of smooth muscle bundles helped to rule out myxoid variant of stromal tumor and myxoid leiomyoma, respectively.

In our case, considering the location of lesion, two close differential diagnoses were superficial angiomyxoma and angiomyofibroblastoma of cervix, which were ruled out histologically. Superficial angiomyxomas arise most often in the head and neck region and occasionally in the vulvovaginal region. They are slowly growing and circumscribed nodules. They are usually less than 3-4 cm in diameter. Histologically multiloculated, poorly delimited myxoid mass composed of plump spindle cells, numerous thin walled blood vessels, and inflammatory cells. Thick walled blood vessels are not seen in superficial angiomyxoma. These tumors have potential for local nondestructive recurrence in approximately 30% cases.[6]

Angiomyofibroblastomas are well circumscribed, round, ovoid, or lobulated, usually less than 3 cm in diameter. Majority of them measure 2-8 cm. The cut surface is gray-pink to yellowish-brown to tan. There are hyper and hypocellular areas. Perivascular hypercellularity is present. Epithelioid plump spindle cells, multinucleate cells, and many thin walled blood vessels are present.
Recurrence is rare. Babala et al. reported a case of angiomyxofibroblastoma of the cervix uteri. Recognition of this entity is important to avoid misdiagnosis of the other angiomyxoid neoplasms.

Van Roggen et al. reported a clinicopathological and immunohistochemical study of 11 cases of AAM. All cases arising in females were positive for desmin, while three of the six cases arising in males were negative for desmin. Strong diffuse positivity for CDK4 was observed in all six cases. Six of eleven cases were positive for alpha smooth muscle antigen. Five of eleven were CD34 positive.

Skalova et al. described a rare case of AAM presenting as an endometrial uterine polyp. This was the first case of AAM within the uterine cavity. Complete surgical excision is the gold standard for AAM because of its tendency to recur locally. The recurrence rate varies from 36% to 70%. Surgery causes significant morbidity due to its frequent occurrence in lower pelvis and perineum with proximity to genitourinary and anorectal structures. Most surgeons aim at complete resection (wide excision with tumor free margin), incomplete or partial resection is acceptable when high operative morbidity is anticipated and fertility is an issue. Treatment options include use of hormonal manipulation such as tamoxifen, raloxifen or GnRH analogs, radiotherapy, and arterial embolization..

It is typically a benign, nonmetastasizing neoplasm. In two cases, however, multiple metastasis have been reported. Hence, long-term follow-up of patient is necessary. Our patient is followed up for 6 months after surgery. There is no evidence of recurrence till date.

REFERENCES