International Journal of Science and Research (IJSR)
ISSN (Online): 2319-7064
Index Copernicus Value (2016): 79.57 | Impact Factor (2017): 7.296

Definitive Radiotherapy for Vaginal Angiomyofibroblastoma

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Abstract: ***Angiomyofibroblastoma (AMF) is a rare,benign, mesenchymal tumour occurring mainly in the vulva region of women. These tumours occur primarily on vagina, vulva, perineum, uterine cervix and the inguinoscrotal regions of men. There are only few cases described with primary occurrence in the pelvis or retroperitoneum. The goal of management is complete resection of tumour, but incomplete or partial resection is acceptable, especially when high operative morbidity due to extensive surgery is anticipated and preservation of fertility is an issue.Most of these tumours show oestrogen and progesterone receptor positivity and are likely to be hormone dependent. Several beneficial results with gonadotropin-releasing hormone (GnRH) agonist have been described in primary treatment of small tumours, as adjuvant therapy for residual tumour, pre-operatively to shrink tumours or even in the treatment of recurrence. However, there are no conclusive data as to theeffectiveness of hormonal therapy/ oopherectectomy in the treatment of these tumours. There are no supporting data for use of chemotherapy in patients with AMF.Radiotherapy may be good alternative to surgery; for unresectable, medically inoperable or recurrent tumours, and also patients unresponsive to embolization or hormonal therapy requiring morbid surgery. This case report describes the unique role of definitive radiotherapy for recurrent unresectableangiomyofibroblastoma of vagina in a young woman; and remains complete response for 20 months.We emphasize the role of definitive radiotherapy for patients with angiomyofibroblastoma of vagina and other pelvic sites that are not amenable for surgery, with benefits of good local control and organ preservation.***

Keywords: Angiomyofibroblastoma, Vagina, Radiotherapy

1. Introduction

Angiomyofibroblastoma(AMF) is a rare and benign soft tissue tumour first described by Fletcher et al. in 1992. It belongs to the group of genital mesenchymal tumors mostly occurring in lower genital tracts of young to middle-aged women (1). These tumours occur primarily on vagina, vulva, perineum and uterine cervix (2-4) and the inguinoscrotal regions of men (5). There are only few cases described with primary occurrence in the pelvis or retroperitoneum (6,7). It should be differentiated from aggressive angiomiyxoma, cellular angiofibroma, and other myxoid tumors of genital tract (8), in which radical surgical treatment is needed.We report a case of young post hysterectomisedwomen with recurrent unresectable aggressive angiomyofibroblastoma treated with primary radiotherapy.

2. Case Report

A 29 years old female with para 2 and living 2 presented to department of oncology, Vydehi Institute of Medical Sciences and Research Center, Bengaluru with complains of mass and bleeding per vagina since 5 months in Jan, 2016. There was no history of pain abdomen and abdominal distension/ altered bowel or bladder movements or loss of weight appetite. She had no associated comorbidities either.She had undergone hysterectomy for uterine fibroid in Jan 2014. Theper vagina examination showed about 5x5x5cm sized mobile polypoidal growth seen in the right lateral and posterior wall of vagina till introitus.The excixion biopsy was reported as Angiomyofibroblastoma of vagina. Patient had undergone wide local excision of the tumour and follow up was advised.

Later in May 2016, she again presented with mass per vagina and bleeding.The per vaginal examination revealed a recurred firm growth measuring 5x5x7cm on the right lateral vaginal wall extending till the vault, which bleeds on touch.The new histopathology of the biopsied growth revealed alternating hypercellular and hypocellular edematous regions with abundant blood vessels. There is minimalnuclear atypicality. The immunohistochemistry reported strongly positive for actin, vimentin, CD34, ER/PR, and negative for desmin; suggestive of angiofibroblastoma. Magnetic Resonance Imaging (MRI) of pelvis revealed post hysterectomy status with an ill-defined heterogeneously enhancing polypoidalT2 hyperintense lesion of size 45x43x86mm involving the vaginal vault and inferiorly the lesion is seen involving the vulva. Multiple sub-centimetric left iliac and B/L inguinal lymphnodes noted.

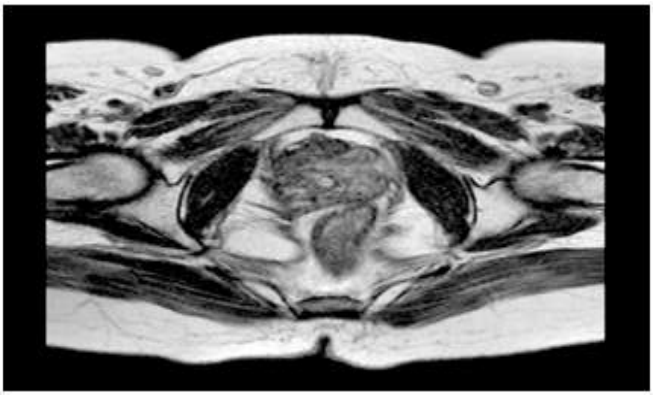

Figure 1: Heterogeneously enhancing polypoidal T2 hyperintense lesion of size 45x43x86mm involving the vaginal vault and inferiorly the lesion is seen till introitus

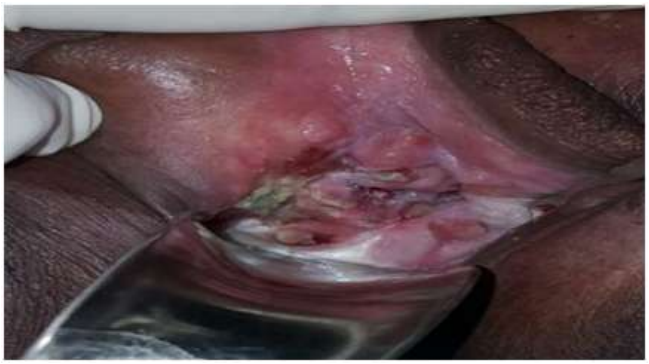

Figure 2: 5x5x5cm sized mobile polypoidal growth seen in the right lateral and posterior wall of vagina till introitus

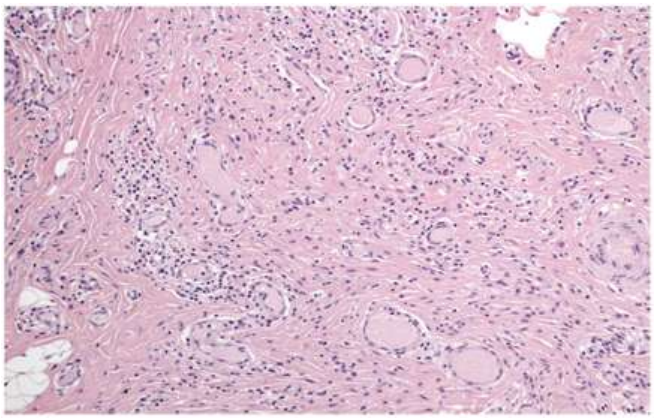

Figure 3: Mixture of hypercellular and hypocellularedematous areas with abundant small- to medium-sized vessels

Our multidisciplinary board decided to treat AMF of vagina with primary radiotherapy in view of aggressive recurrent unresectable disease. Patient received external beam radiation to local diseased area with margins to a dose of 50Gy in 25 fractionfollowed by boost dose of 30Gyin 5 fractions of interstitial brachytherapy from 24/May to 20/July/2016. Patient tolerated treatment well and during course of radiotherapy, she had developed skin grade II toxicities which were managed medically. Patient was kept on close follow up with clinical examinationand remains with complete response for 20months**.**

3. Discussion

A wide variety of mesenchymal lesions occur in the lower female genital tract (9,10). Broadly, these mesenchymal lesions can be separated into two groups. The first group includes several well characterized tumors that show a marked tendency to occur in the lower female genital tract, such as aggressive angiomyxoma, angiomyofibroblastoma (AMF) and cellularangiofibroma (11). These tumours can also be called as relatively site-specific. The second group embraces a wide range of heterogeneous lesions that frequently occur in this region, but arise in other anatomic sites as well, with examples, superficial cervicovaginalmyofibroblastoma characterised by its numerous small- to medium-sized thick-walled vessels and negative staining for desmin. Other lesions that may be confused with SCVM include solitary fibrous tumour and mammary-type myofibroblastoma.Angio-myofibroblastoma of the lower female genital tract is diagnosed based on clinical, radiological and morphological patterns treated primarily with surgery. Adjuvant therapy with radiotherapy in invasive forms may be recommended (12).

One should aim for complete resection, but incomplete or partial resection is acceptable, especially when high operative morbidity due to extensive surgery is anticipated and preservation of fertility is an issue. Long-term follow-up and careful monitoring with imaging techniques are essential for timely identification of recurrence. However, except for positive surgical margins, there are no clinical or histological predictors for tumour recurrence(13,14). The data in literature with limited number of patients with vaginal AMF reports 1-2year of recurrence free survival (1,15).Most of these tumours show oestrogen and progesterone receptor positivity and are likely to be hormone dependent. Several beneficial results with gonadotropin-releasing hormone (GnRH) agonist have been described in primary treatment of small tumours, as adjuvant therapy for residual tumour, pre-operatively to shrink tumours oreven in the treatment of recurrence(16,17). However, there are no conclusive data as to theeffectiveness of hormonal therapy/ oopherectectomy in the treatment of these tumours(18).Due to low mitotic activity, radiotherapy or chemotherapy is unlikely to be a useful adjunct to primary surgery. Most authors did not notice any advantage of radiation therapy(19,20). Pre-operative external beam irradiation and intra-operative electron beam radiotherapy was used in one case to reduce risk of recurrence but follow-updata on the same are not available. Two cases of successful control of recurrent angiomyxoma with relatively high doses of external radiotherapy have also been reported (21,22). There are no supporting data for use of chemotherapy in patients with AMF.

Radiotherapy may be good alternative to surgery; for unresectable, medically inoperable or recurrent tumours, and also patients unresponsive to embolization or hormonal therapy requiring morbid surgery(23).This case report describes the unique role of definitive radiotherapy for recurrent unresectable angiomyofibroblastoma of vagina in a young woman; and remains complete response for 20 months.

4. Conclusions

We emphasize the role of definitive radiotherapy for patients with angiomyofibroblastoma of vagina and other pelvic sites that are not amenable for surgery,with benefits of good local control and organ preservation.

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