Aggressive Angiomyxoma of Labia Majora-A Case Report and Literature Review

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Abstract

We present here a rare case of aggressive angiomyxoma in labia majora. Routine hematological investigations were done. The patient like many villagers was non cooperative for more investigations, but agreed for excision to get permanent cure. Mass was removed. Histopathological report confirmed the diagnosis. Two years follow up did not show recurrence.

Keywords: Aggressive angiomyxoma; Labia majora; Female; Vulva

Introduction

Aggressive Angiomyxoma (AAM) was first described by Steeper and Rosai [1]. The tumor was named aggressive due to its characteristically slow and insidious growth as well as carrying a high risk of local relapse. AAM emphasizes the neoplastic nature of the blood vessels and its locally infiltrative and recurrent nature. It is a rare local mesenchymal tumor of unknown etiology usually affecting vulva, perinial region, buttocks or pelvis of reproductive age [2-6]. Fewer than 250 cases have been reported till 2010 [7]. Few cases were reported in adult male and children (age group 8-13 years) [8,9]. The ratio of female to male cases was 6:1 [10]. Clinical presentation mimics a Bartholin’s cyst, lipoma, perineal hernia or pedunculated soft tissue tumor in females, while in males it commonly presents similar to hydrocele or inguinal hernia [11,12]. AAM might get diagnosed as benign or malignant lipomatosum tumor when this entraps adipose tissue. While growing if not treated an AAM would displace and invade other organs.

Case Presentation

Thirty nine year old para 2 presented with vulval swelling on the right side with duration of 3 years and increase in size since 6 months (Figure 1). There was no history of vulval discharge, pain, bleeding or dyspareunia. Bowel and bladder habits were normal. Menstrual cycles were regular with normal flow. Local examination showed a non tender, non reducible, non compressible pyriform swelling on the right side with duration of 3 years and increase in size since 6 months. Skin over the swelling appeared normal. Detailed gynaecological examination revealed uterus in normal size and no change in well developed secondary sexual characteristics. There was no lymphadenopathy.

Routine blood investigations were done and results were within normal range. Other investigations including fine needle biopsy were not done due to non cooperation of patient. Surgical intervention was agreeable to the patient to eliminate discomfort.

Excision of the mass was done under spinal anaesthesia (Figure 2). Histopathology of the excised specimen was suggestive of angiomyxoma. In the case reported here tumor weighed 400 gm. On gross examination the tumor was seen lobulated at areas, other areas with poor demarcation and soft to rubbery in consistency. It was bulky having tan grey colour. The cut surface revealed a glistening, gelatinous and soft homogeneous appearance. Congested blood vessels with occasional hemorrhage were seen. Histopathology results showed spindle shaped cells with abundant eosinophilic cytoplasm and blood vessels of varying calibre scattered in a myxoid background. The tumor showed high content of water. There was no atypia or mitosis. Follow up was done for two years period showing no recurrence.

Discussion

The tumor, AAM commonly presented as an asymptomatic mass in the genital area. The differential diagnosis of this unusual tumor AAM includes myxoma, myloid lysosarcoma, sarcomabotyoids, myxoid variant of malignant fibrous histioctyoma, angiomyofibrolastoma, rhabdomysarcoma, nerve sheath myxoma and other soft tissue tumors with secodary myxoid changes [1]. Many times it was confused for a gynaecological malignancy, due to the rarity of the tumor. Zhang [13] reported one case of AAM with massive asicites. It is a rare and slowly growing, tumor with a less propensity to metastasize. Metastatic AAM was reported by Geng et al., Blandamura et al. and Siasii et al. [14-16]. High rate of angiomyxomas were reported in skeltal muscle [17]. Rare cases of angiomyxoma were seen in larynx [18], lung [19], maxillary sinus [20], jejunum [21] and urinary bladder [22]. In males, commonly seen sites are scrotum, inguinal region and perineum. Rare cases involving the epididymus [23] and spermatic cord [24] were also reported. Due to its highly infiltrating nature, AAM had a high tendency for recurrence; though Rosai [17] opined less recurrence majority of cases. The recurrence rate has been reported as 70% and seen in 2 years period, though cases of recurrence were reported after 8...
years [25] or even after 20 years [26,27]. It made necessary to follow up all cases after excision for minimum 2 years period.

Size of the neoplasm varied from 5 to 23 cm. Surgery by margin free excision was the mainstay of treatment. Laparoscopic removal of angiomyxoma was not uncommon [28]. Prior to surgery misdiagnosis of AAM was not rare due to its rarity. Güngör [29] reported its rate as 82%. AAM is generally diagnosed histologically [7].

The tumor cells were immunoreactive to vimentin, desmin and smooth muscle actin but negative to S100 protein [30]. Most tumors exhibit estrogen and progesterone receptor positivity in females [31]. This may be suggestive of a role of hormones supporting this a few reports showing rapidly growing tumor during pregnancy [32]. During pregnancy progesterone receptors were strongly positive whereas estrogen was negative [32-35]. Resected specimens from male patients were positive for androgen and progesterone receptors [36]. Cytogenetic analysis showed abnormal chromosomal translocation12:21 [46, XX, t(12;21)(q15;q21.1)] and rearrangement of HMGA2 gene [20]. HMGA2 was positive in most aggressive angiomyxomas and was useful in diagnosis as most mesenchymal lesions which closely mimic this are negative.

Preoperative angiographic embolization, preoperative external beam irradiation and intra operative electron beam radiotherapy were useful to decrease the chances of local recurrence [37], as well as being of value in primary diagnosis; HMGA2 was useful in evaluating margins and in reexcision specimens of aggressive angiomyxomas in identifying foci of residual or recurrent tumor. Hormonal treatment with GnRHa analogues were successfully used to treat recurrence in few cases and as primary therapy for small tumors [33,38].

References


