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AGGRESSIVE ANGIOMYXOMA OF THE PERINEUM ORIGINATING FROM THE RECTAL WALL: A CASE REPORT

Oncology

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KEYWORDS

INTRODUCTION

Aggressive angiomyxoma (AA) is a rare soft-tissue tumor characterized by its locally infiltrative behavior. AA is an uncommon mesenchymal tumor which is predominantly encountered among adult females in reproductive age[1]. The tumor usually arises from the pelvic and perineal regions; however, uncommon localization has been reported in the literature[4,5]. AA is a locally infiltrative slow growing tumor with a marked tendency to local recurrence. Although it is previously regarded as a nonmetastasizing tumor, its metastatic potential has been revealed in a few recent reports. Macroscopically, AA has a gelatinous appearance, and it is microscopically characterized by a myxoid stroma and abundant thin-thick walled vascular channels. The tumor is distinguished from other lesions by these Histopathological features. The most common clinical symptom is painless swelling at vulva or groin area. Because of this clinical manifestation, AA is often initially misdiagnosed as a gynecological malignancy or a groin hernia that leads to unnecessary surgical interventions. Complete surgical excision with tumor-free margins is still accepted as the main treatment method for AA. Unfortunately, the highest recurrence rates after resection still remain a major surgical problem that should be solved. Since AA was first described in 1983 by Steeper and Rosai, approximately 200 cases have been reported in medical literature up to date. Because of its rarity, the clinical presentation and the treatment method of the tumor have been described mostly based on individual case presentations.

It is noteworthy that there is still a lack of knowledge about the clinical presentation, the management options, and the follow-up results of AA in the current literature. Hence, we aimed to contribute our case to the literature by presenting the surgical outcome of a patient who underwent radical surgery for an AA.

Case report

A 32 year-old woman was admitted to our hospital with a mass in the perineal region, associated with mass protruding per vaginum. She was otherwise healthy, with no remarkable past medical history or family history. She underwent tubectomy in past. On physical examination, a vague swelling just under the left gluteal cleft of size around 4x6 cms. Expansile swelling present in vagina of size around 6x6cms.Margins were indistinct and swelling was compressible. Cough impulse was positive in the swelling. Computed tomography scan revealed a large lobulated heterogeneously enhancing predominantly cystic lesion in pelvis along posterior aspect of uterus. The lesion is compressing and right laterally displacing the anorectum. Lesion also extending in perineal region on left side. In addition, magnetic resonance imaging showed the mass originating from rectal serosa, extending from S3 vertebral body to subcutaneous region of the perianal region on the left side. Posteriorly mass occupies the presacral space. No intralesional calcification or necrosis with maintained fat planes. Incisional biopsy was obtained by a perineal approach showing non neoplastic colonic mucosa, mild lymphoplasmacytic cell in stroma.malignant cells were not seen. Colonoscopy revealed thickened ano rectal mucosa and esophagogastroduodenoscopy was normal.Pre-operative CEA, CA125, CA19-9 was.92 ng/ml; 11.69U/ml; 9.53U/ml respectively. Pap smear was negative for intraepithelial malignancy. Complete resection of mass was done by abdominal approach. Histopathological examination post operatively revealed macroscopically two soft tissue masses, one measuring 13 x 7 x 2.5 cm, other 9.5 x 5.6 x 4 cm. external surface glistening, and smooth, gray white. Cut section- capsulated grayish white solid. Microscopically the sections show capsulated tumor composed of spindle cells forming loose sheets in a dense myxoid stroma with proliferation of small blood vessels. Tumor cells forming whorling pattern surrounding vessels suggesting it to be an AA. The patient recovered uneventfully and there has not been any evidence of local recurrence for 2 months postoperatively.

DISCUSSION

AA is an uncommon mesenchymal tumor which is mostly derived from the pelvic and perineal regions including vulva, vagina, bladder, and rectum[1,2,3]. However, uncommon localizations such as lung, liver, larynx, and orbit have been reported[4,5,6,7]. It is hard to define the exact incidence of AA among the other intra-abdominal mesenchymal tumors because of its rarity. Although it is almost exclusively encountered among females in reproductive age, rare cases have been diagnosed in the perimenopausal female, children, and male patients[8]. The female-to-male ratio has been reported as 6.6/1[9]. The reported age of presentation ranges from 1 to 82 years, with a median age of 33 years. In view of these data, the demographic characteristic and the tumor localization of our patient were similar to the majority of previously reported cases. The pathogenesis of AA is not well understood. There is only one study evaluating the pathogenesis of AA in the current literature in which Nucci and Fletcher[10] suggested a translocation at the level of chromosome 12 where the high mobility group protein HMGA2 (a transcription factor expressed during embryogenesis) is located. AA is regarded as an aggressive tumor due to the neoplastic nature of the blood vessels and its high tendency of local infiltration and local recurrence. AA is considered as an invariably benign tumor; however, in a few recent reports, its metastasis to lungs resulting in death has been revealed. This condition which highlights the need to consider AA may be potentially malignant in some cases[4]. AA should be distinguished from other relatively more common encountered benign or malignant soft-tissue sarcomas of the abdomen such as myxoma, fibrous histiocytoma, angiofibroma, liposarcoma, nerve sheath tumor, mixed mesodermal tumor, and angiomyofibroblastoma[1,2]. AA is distinguished from the other lesions by its immunohistological findings. AA is derived from myofibroblasts as a phenotypic variant of the basic fibroblast with a prominent vascular component[6] .The origin of the tumor may act like a wound healing situation and this may be the reason of its locally invasive character. Immunohistochemical staining of the tumor reveals high positivity for desmin, vimentin, ER, and PR receptor; however it generally reveals negativity for S-100 protein[11]. These findings usually help us to distinguish from other mesodermal origin tumors. The diagnosis of AA is very difficult because it is often asymptomatic until the tumor reaches large sizes. Urinary, gynecologic, and gastrointestinal symptoms like dysuria, dysmenorrhoea, constipation, and chronic abdominal/pelvic pain occur when the tumor begins to depress the adjacent organs including bladder, rectum, ureter, and uterus. AA commonly manifests a painless swelling located around the genitofemoral region. For this reason, it is often misdiagnosed as a vulvar abscess, Bartholin's gland cyst, vaginal prolapse, gynecologic malignancy, and groin/femoral hernia that may lead to unnecessary surgical interventions. Furthermore, because the size of the tumor is often underestimated by clinical examination, these imaging studies also help us in deciding the surgical strategy. In view of these findings,

we think that there is still a lack of knowledge about the diagnosis of AA among the clinicians. Thus, we emphasize that although it is a rare condition, particularly, a female in reproductive age with a painless swelling located around the genitofemoral region should be well examined by imaging studies to rule out AA and to decide the surgical strategy, as well.

The current treatment of AA is complete surgical excision with tumor-free margins[1,3,6]. However, there is still a debate about the treatment because of high recurrence rates in spite of wide surgical excision. The recurrence rate is reported with a wide range from 33% to 83%[12]. Recurrence has been developed mostly within the first 3 years[13]. However, it may be detected even after postoperative 15 years. In a retrospective review by Chan et al.[13] and Han-Geurts et al.[14], it has been suggested that patients having positive margins were as likely to have recurrence as those with negative margins. Also, the tumor size is not correlated with recurrence. Thus, extensive surgery can be disregarded in patients with high morbidity and for preserving fertility, as well. Respecting this view, we believe that it is not compatible with the oncological principles. Incomplete or partial resection may lead to high recurrence rates. Furthermore, considering the recently reported cases with distant metastasis, complete resection should be performed as technical as possible despite the high morbidity of the operation.

All adjuvant treatment modalities remain controversial[8]. Chemotherapy yields no beneficial results for adjuvant therapy because of low mitotic activity of the tumor. Embolization of the tumor has been reported as an alternative approach; however, it remains insufficient due to the extensive vascular network of the tumor. The main localization of AA, which is limited to reproductive organ region, and the positive ER and PR status of the tumor suggest that AA may be a hormone-responsive neoplasm[1]. Several beneficial results with tamoxifen or gonadotropin-releasing hormone agonist have been described[1,9]. Fine et al. [15] achieved a complete resolution of a recurrent AA in a female patient who refuses redo surgery. However, long term use of these drugs is associated with side effects such as menopausal symptoms and bone loss.Moreover, the optimal duration of therapy is unknown.The immunohistochemical findings of the present tumor confirmed positivity for both estrogen and progesterone receptors.Although the majority of the authors have reported no advantage in using radiotherapy, it can be a good alternative treatment in patients who are resistant to antihormonal therapy, those with recurrence or in whom tumor resection would cause high morbidity. Rhomberg et al.[16] and Suleiman et al.[17] have achieved local control by radiotherapy in patients with local recurrence.

CONCLUSION

Despite its rarity, AA should be considered in the differential diagnosis of any painless swelling located in the genitofemoral region, particularly in women of reproductive age. The diagnosis should be confirmed by CT and/or MRI. The principle treatment should be complete surgical excision with tumor-free margins. The patient should be informed about the high morbidity of the surgical intervention. Long-term follow-up and careful monitoring are essential due to its high tendency of local recurrence in spite of wide excision of the tumor. Adjuvant antihormonal therapy yields promising results for preventing recurrence. However, long-term use of these drugs is still controversial because of their adverse effects.

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