Journal of Cancerology. 2014;1:80-3

A Case Report of Angiomyxoma with Complete Response to Goserelin plus Tamoxifen: Review and Update

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ABSTRACT

Aggressive angiomyxoma is a rare mesenchymal tumor with a locally infiltrative growth pattern. It preferentially involves the pelvic and perennial regions. It is a locally infiltrative neoplasm with a marked tendency to recurrence, although distant metastases are extremely rare. Radical surgical resection is considered the mainstay of treatment. However, the risk of recurrence is high, so therefore multimodal treatment is needed. Knowing that most of these tumors are estrogen and progesterone receptor positive, it has been hypothesized that they may respond to hormone treatment. We report the case of a 34-year-old woman with a tumor in right gluteal region. The biopsy revealed aggressive angiomyxoma. The patient was treated with surgery. However, 3 months later she developed locoregional recurrence. We started systemic hormonal treatment with tamoxifen and goserelin. After 14 months of treatment there was partial response. (J CANCEROL. 2014;1:80-3)

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Key words: Angiomyxoma. CT-guided biopsy. Vimentin. Positive for desmin. ER and PR.

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Received for publication: 30-10-2014
Accepted for publication: 18-11-2014

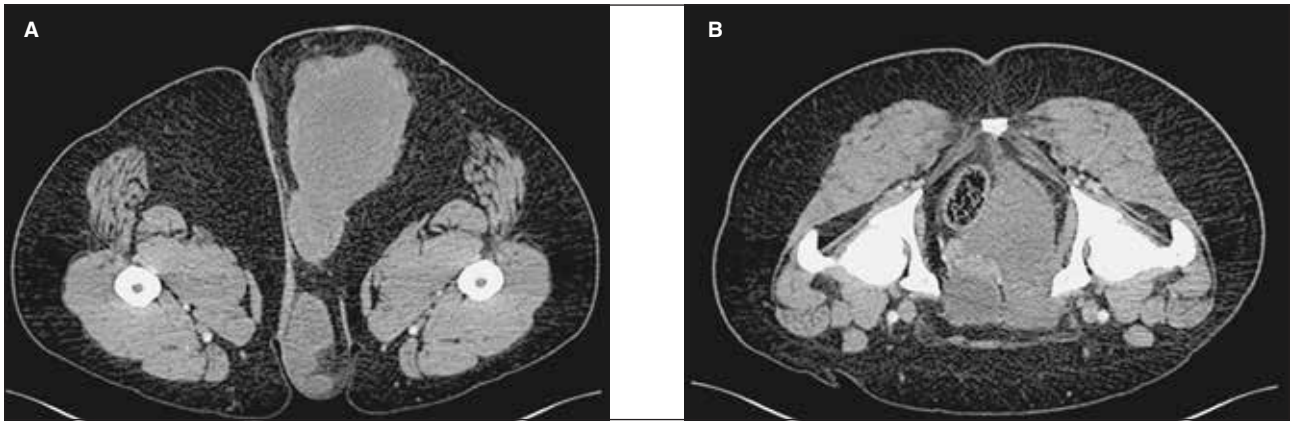

Figure 1. 1A. CT scan showing intra- and extra-pelvic mass. **1B.** CT scan showing locoregional recurrence.

INTRODUCTION

Aggressive angiomyxoma is a rare mesenchymal tumor with a locally infiltrative growth pattern. It preferentially involves the pelvic and perineal regions. The low-grade form is more frequent in women than in men by a ratio of 6:1 and most commonly occurs in women of reproductive age[1,2].

Radical surgical resection is considered the mainstay of treatment. However, the risk of recurrence is high, so therefore multimodal treatment consisting of radiotherapy, hormone therapy and angiography was offered, although there is little experience in this regard[1,2]. We report the case of a 34-year-old woman with a 20 × 15 cm tumor in right gluteal region evolving since two years and occasional pain controlled with nonsteroidal analgesics. A computerized tomography (CT)-guided biopsy revealed: aggressive angiomyosarcoma, positive for vimentin and desmin, negative for CD34, positive for estrogen receptor (ER) and progesterone receptor (PR). In November 2012 extensive surgery was performed and three months later locoregional recurrence was observed, so in 2013 systemic hormonal treatment with tamoxifen and goserelin administered orally was decided on. After 14 months of treatment there was partial response.

CASE REPORT

A 34-year-old woman whose symptoms began with the appearance of an insidiously growing tumor in the right gluteal region evolving since two years and associated with occasional pain controlled with nonsteroidal analgesics.

Her physical examination upon admission revealed a 20 × 15 cm ovoid tumor in the right gluteal region, with well-defined edges, soft consistency, extending to the perineum and right labia majora. The CT scan showed a large intra- and extra-pelvic mass occupying the pelvic cavity, right presacral space, and gluteal region, with extrinsic compression of the rectum, bladder, and uterus (Fig. 1 A). Routine blood count, blood chemistry, and liver function tests were normal.

We decided to perform a CT-guided biopsy, which revealed: aggressive angiomyosarcoma, positive for vimentin and desmin, negative for CD34, positive for ER and PR. With these data, the patient underwent an extensive lumpectomy in November 2012. The surgical findings were: myxoid tumor with a rubbery consistency and cystic areas, extending from the vulva to cover most of the right buttock and extending into the pelvic cavity, of approximately 35 × 20 × 15 cm. The histopathological

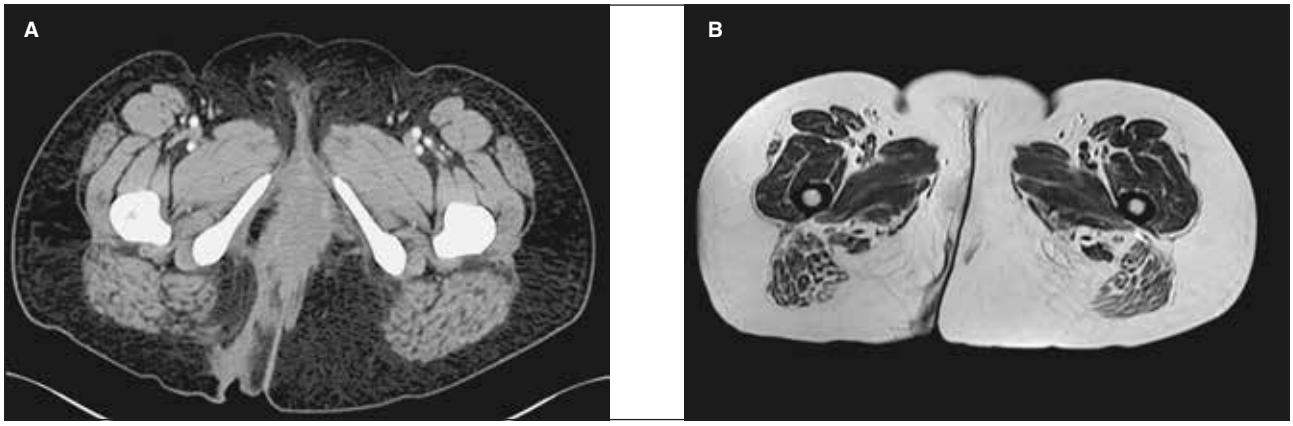

Figure 2. 2A. CT scan showing neoformation. **2B.** MRI showing complete response.

study showed: aggressive angioma with a 15 cm maximum diameter, with tumor pushing surgical boundaries.

In the postoperative period she developed a fistula between the rectum and the gluteus, so transverse loop colostomy was performed. Three months after surgery she had locoregional recurrence with a tumor measuring 80 × 60 mm (Fig. 1 B), so she was sent to medical oncology for systemic therapy treatment in February 2013. Treatment was initiated with goserelin 10.8 mg monthly and tamoxifen 20 mg orally daily.

After nine months of treatment, the CT scan showed a neoformation measuring 25 × 22 × 27 cm (Fig. 2 A). With this information, we concluded partial response had been achieved, so the abovementioned treatment was continued. To date she has been in treatment for 14 months with goserelin and tamoxifen, with clinical data showing no tumor activity and control magnetic resonance imaging (MRI) showing complete response (Fig. 2 B).

PATHOLOGICAL FINDINGS

Pathological analysis of the tissue resected from the right buttock consisted of a multinodular mass with soft consistency, irregular borders, formed by tissue that had a striated muscle appearance and areas with a café-au-lait myxoid appearance; its longest axial diameters measured 10.5 × 8.5 × 3.5 cm. The cut surface of this tumor is café-au-lait, with a myxoid appearance and can be seen infiltrating the striated muscle tissue. Macroscopically, the tumor tissue is at the resection borders.

Histologically, the tumor showed mesenchymal features with lax areas, the cells were fusiform with focal areas of polygonal cells, with scarce cytoplasm, basophilic nuclei; the vascular capillary network with slightly thickened walls has the classic features of aggressive angiomyxoma.

Immunohistochemically, the tumor was positive for desmin, vimentin, and hormone receptor-positive for estrogen and progesterone.

DISCUSSION

Aggressive angiomyxoma is a mesenchymal neoplasm described by Rosai and Steeper in 1983. It affects the deep soft tissues of the vulvovaginal region, perineum, and pelvis of women of reproductive age[3]. It is a locally infiltrative neoplasm with a marked tendency for recurrence, although distant metastases are extremely rare[3].

Recurrence is common in approximately 30-40% of cases and may occur from two months to 15 years later[6]. Surgical excision is usually the first line of treatment for both primary tumors and tumor recurrences, although it results in high morbidity and aggressiveness. Other treatment modalities have been described, such as hormone therapy, radiation therapy, and angiography.

Knowing that most of these tumors are ER and PR positive, it has been hypothesized that they may respond to hormone treatment[2].

There have been few studies in this area, some with gonadotropin-releasing hormone (GnRH) agonists given as monotherapy, resulting in complete remission of the recurrence[3] and others resulting in a marked decrease in size of the tumor[4,5]. Hormonal manipulation with other inhibitors such as tamoxifen or raloxifene has been tried in a few studies, but their role is not clearly defined[6].

The first report of the use of GnRH agonists was published by Fine, et al. following treatment of a 34-year-old patient who had had two prior surgical excisions for recurrences, to whom GnRH agonists were administered for six months, achieving a complete response for more than six months[1]. Likewise, McCluggage, et al. describe the dramatic response of aggressive angiomyxoma of the vulva to therapy with a GnRH agonist in a 35-year-old patient who had an extensive tumor mass on the vulva that was invading the center of the pelvis, who underwent major surgery and subsequently received 3.6 mg of Zoladex® (goserelin) monthly for eight months to treat the residual tumor and in whom pathologic complete response was documented[3].

In our case, we decided on a double hormonal blockade with Zoladex®, a synthetic analogue of GnRH. Chronic administration of goserelin causes the inhibition of the secretion of pituitary follicle stimulating and luteinizing hormone, leading to a state of hypoestrogenism[3].

On the other hand, tamoxifen is a selective ER modulator. Its mechanism of action is based on its antiestrogenic effects, i.e. it blocks the action of the hormone that stimulates the growth of tumor cells. It is not useful for every cancer, only for those whose cells have specific receptors for estrogens. It is commonly used to treat premenopausal breast cancer patients with ER-positive cancer[7]. However, the use of tamoxifen in patients with aggressive angiomyxoma has not been documented.

In our patient we decided on a double hormonal blockade using a GnRH agonist and tamoxifen, with excellent results and virtually no adverse effects to treatment.

CONCLUSION

We report the case of a patient with angiomyxoma in the right buttock extending into the pelvis, with recurrence after primary surgical treatment with radiological complete response to administration of GnRH agonists and tamoxifen. This combination may be considered in the management of recurrent aggressive angiomyxoma with hormone receptor positivity, with excellent response and minimal side effects.

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