International Journal of Medical and Health Research
ISSN: 2454-9142
www.medicalsciencejournal.com
Volume 1; Issue 5; December 2015; Page No. 28-29

Aggressive angiomyxoma of cervix presented as huge broad ligament tumour: A case report

[1] Dr. Shamim Khandaker, [2] Dr. Shabana Munshi

[1] Assistant Professor, North Bengal Medical College, Sushrutanagar, Darjeeling, India.

[2] R.M.O cum clinical tutor, North Bengal Medical College, Sushrutanagar, Darjeeling, India.

Abstract

Aggressive angiomyxoma is a rare, slow growing, benign soft tissue tumour of pelvis and perineum. We presents a case of aggressive angiomyxoma of cervix presented as one fairly large (77.2x60.9x76.2 mm^3) echogenic heterogeneous space occupying lesion in left adnexa mimicking broad ligament fibroid. It should be distinguished from various others soft tissue tumours in this region due to various recurrence potential of these tumours. Wide local excision with tumour free margin is the treatment of choice along with long term follow-up and careful monitoring with imaging techniques; which are necessary before the possibility of a recurrence can reasonably be dismissed.

Keywords: Aggressive angiomyxoma; Pelvic soft tissue tumour

Introduction

Aggressive angiomyxoma is a rare, slow growing, benign soft tissue tumour of pelvis and perineum. This was first described in 1983 by Steeper *et al.* [1] This is a mesenchymal tumour arises from connective tissue of lower pelvis or perineum and it has a locally aggressive course and can be recurrent [1]. It is important to diagnose this condition because the tumour is locally infiltrative and requires wide excision and follow up.

Case report

A 45 years old postmenopausal woman was presenting with vague abdominal pain and mild abdominal swelling for last 6 months. On per vaginal examination, uterus was found normal in size with restricted mobility and an approximately 6x7 cm firm mass found in left adnexa. On transvaginal ultrasonography (TVS); uterus was retroverted and sized 8x3.7x4.9 cm^3, bilateral ovary was normal in size, along with one fairly large (77.2x60.9x76.2 mm^3) echogenic heterogeneous space occupying lesion (SOL) in left adnexa and POD region and another similar SOL (46.3x17.5x36.8mm^3) in right adnexa. Her blood CA-125 level was 21.10, which was normal. We planned for laparotomy, where the SOL was found to be originating from lower segment of uterus extending towards the broad ligament. We performed total abdominal hysterectomy and bilateral salpingo-oophorectomy along with complete removal of the SOL en-mass (Figure 1). Grossly the SOL was a 9x7x3cm globular cyst arising from upper outer wall of uterus; uterus size was 10x6x4.5cm and both the ovaries were 2x2cm. Cut surface of the SOL showed peripheral cystic areas around central solid homogeneous areas. The specimen was sent for histopathology. The section showed feature of hypo-cellular areas with myxoid changes having proliferation of fibroblast and myofibroblast; there was proliferation of medium and small sized blood vessels with no cellular atypia or mitosis; suggestive of aggressive angiomyxoma of cervix and broad ligament.

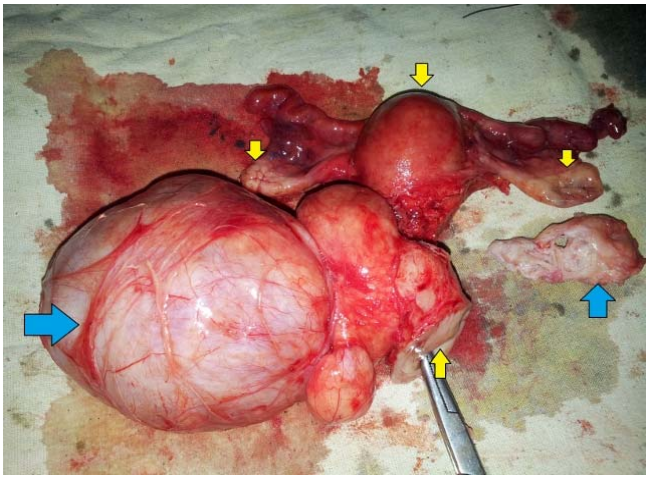

Fig 1: Uterus and bilateral fallopian tubes and ovaries (yellow arrows) with cervix (artery forceps) and Aggressive angiomyxoma of cervix (blue arrows)

Discussion

Aggressive angiomyxoma derives from a primitive mesenchymal cell normally found in the soft tissue of pelvis, which is capable of myofibroblastic differentiation. It is usually present as growth/ mass arising from the vulva or perineum. Very rarely, it has presented as a cervical polyp or a uterine polyp where it presented with uterine bleeding [2]. Our case is unique in the sense that it arises from cervix and grows towards broad ligament bilaterally. This type of growth is confusing to be degenerating fibroid of broad ligament due to solid and cystic nature of the angiomyxoma.

Age distribution is from 6-60 years with peak incidence at 31-35 years. Patients are often asymptomatic and the presenting features are nonspecific such as a palpable mass with heaviness or discomfort in the lower abdomen. Compression to urinary/intestinal systems occurs only when the tumour is large. These tumours are slowly growing and painless [3].

An accurate diagnosis of aggressive angiomyxoma is based on histopathology evidence. Grossly these tumours have a smooth surface with homogenous consistency and a glistening gelatinous appearance on cut section. Microscopy shows a hypocellular mesenchymal lesion with scattered spindled and stellate cells in a myxoid stroma with proliferation of medium and small sized blood vessels along with proliferation of fibroblast and myofibroblast. The tumour lacks cytologic atypia, increased/ atypical mitoses and coagulative necrosis. The tumour is usually positive for vimentin, desmin and negative for S-100 on immunohistochemistry [4].

Aggressive angiomyxoma needs to be differentiated from various others soft tissue tumours in this region which are angiomyofibroblastoma, cellular angiofibroma, superficial angiomyxoma, myxofibrosarcoma and myxoid liposarcoma due to various recurrence potential of these tumours which necessitates different management and follow up options [5].

In case of aggressive angiomyxoma, complete surgical excision (wide excision with tumour free margin) is the treatment of choice, because of its tendency to recur locally. Pre-operative knowledge of tumour extent is important in determining surgical approach [6]. The recurrence rate may be upto 70% which usually occurs within first 3 years; but there is no correlation between size of tumour and recurrence rate. Incomplete or partial resection is acceptable when high operative morbidity is anticipated due to close proximity of genitourinary or anorectal structures or preservation of fertility is an issue. Adjuvant therapy in the form of GnRH analogue, tamoxifen, raloxifene or radiotherapy in cases of incomplete resection and recurrence need to be evaluated [7].

Conflicts of interests: None

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