Case 14535

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A case of recurrent aggressive

angiomyxoma

Published on 04.04.2017

DOI: 10.1594/EURORAD/CASE.14535 ISSN: 1563-4086 Section: Uroradiology & genital male imaging Area of Interest: Abdomen Procedure: Diagnostic procedure Imaging Technique: Ultrasound Imaging Technique: CT Special Focus: Neoplasia Case Type: Clinical Cases Authors: Deshpande Amit A1, Lakhani Nisha2, Shah Dipali3 Patient: 35 years, female

Clinical History:

A 60-year-old female presented with complaints of perineal swelling for 1 year which was gradually increasing in size. The patient was also experiencing difficulty in micturition and defecation for the past week. The patient had undergone surgery for a pelvic mass 10 years before. **Imaging Findings:**

USG showed large hypoechoic solid-cystic lesion extending from the umbilicus to the pelvis. The lesion did not appear to invade surrounding structures. The ureter was seen coursing through the mass.

CECT showed large heterogeneously enhancing solid-cystic lesions extending till the aortic bifurcation superiorly. Inferiorly, the lesion was extending up to the right ischiorectal fossa displacing the rectum and the sigmoid colon without any signs of invasion. There was an extension into the vaginal vault, up to the labia majora. Anteriorly, the lesion was abutting the anterior abdominal wall. The lesion also caused displacement of small bowel loops laterally. Posteriorly, there was compression of the bilateral common iliac arteries by the lesion, without any signs of thrombosis.

There was no sign of invasion into surrounding structures. **Discussion:**

There are few differential diagnoses of perineal soft tissue tumours in adult female patients.

Aggressive angiomyxoma (AAM) is ill-defined, mostly >5cm in size. It is locally invasive and tends to recur, however, distant metastases are not recorded. They grow slowly without invasion of the surrounding structures. [1, 3, 6] Patients present with the complaints of perineal swelling, dysmenorrhoea or dyspareunia.

The true extent is often underestimated on physical examination, because the visible portion usually shows only a fraction of the deep involvement. [5]

CT demonstrates a large locally invasive pelvic tumour, which extends towards the ischiorectal fossa. It may be a well-defined, moderately enhancing mass, hypodense relative to muscle or it may be predominantly cystic with solid components. It is consistent with myxoid matrix, high water content within the mass or both. It does not show gross fat within the mass. [3, 5]

Angiomyofibroblastoma is a localised, well-circumscribed tumour, which usually measures <5 cm in size. It may

show microscopic fat within and does not recur. [2, 4]

Fibroepithelial stromal polyps are relatively common benign lesions of the vulvovaginal region that may be multiple and are associated with pregnancy or exogenous hormone use. Local excision is usually curative. [5]

Myxoid leiomyoma is another pelvic tumour that can grow quite large. Myxoid change may be focal or involve the entire tumour and a helpful diagnostic clue is to identify an area of transition between the myxoid features and moreclassic smooth muscle differentiation histologically. [5]

Differentiating these tumours on the basis of imaging findings is quite difficult as they all share the most imaging features. However, on the basis of clinical history (as in the case of fibroepithelial stromal tumour), the size of the lesion (as in the case of angiomyofibroblastoma) or presence of recurrence, it is possible to reach a conclusion. The final diagnosis is generally made by the biopsy.

The first line of therapy for AAM is surgery, but achieving negative resection margins is difficult because of the infiltrative nature and the absence of a defined capsule. Smaller tumours may be removed with wide, local excision, but larger, deep-seated tumours of the pelvis may require more extensive surgery with partial or complete resection of some pelvic organs. The prognosis is generally good. Recurrence rates range from 25-47% with 85% of those occurring within 5 years of initial surgery. [5, 7]

This patient had undergone surgery for pelvic mass 10 years back which was proven to be AAM histologically and again presented with a similar appearing lesion. Due to the extensive involvement of the lesion, the patient was declared non-operable.

Differential Diagnosis List: Aggressive angiomyxoma, Aggressive yngiomyxoma, Angiomyofibroblastoma, Fibroepithelial stromal polyp, Myxoid leiomyoma

Final Diagnosis: Aggressive angiomyxoma

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Description: Axial CT at the level of the perineum shows the mass lesion extending up to the labia majora. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, India



Description: Axial CT at the level of the bladder shows the mass lesion displacing the rectum and bladder with preserved fat planes. The ureter appears to course through the mass without evidence of invasion. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, India



Description: Axial CT at the level of the perineum shows displacement of perineal muscles due to the mass without definite evidence of invasion. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, India



Description: Axial section just below the level of the aorta bifurcation showing displacement of bowel loops and compression of bilateral common iliac arteries without signs of invasion. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, India



Description: Coronal scan shows the superior and inferior extent of the mass. It displaces the bowel loops laterally without signs of invasion. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, India



Description: Coronal scan shows the mass. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, India



Description: Coronal section shows inferior extent of the lesion with displacement of rectum and perineal muscles without signs of invasion. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, India



Description: USG shows the hypoechoic mass lesion with dilated ureter coursing through the lesion and displacement of bladder with foley's bulb and rectum without signs of invasion. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, Gujarat, India



Description: Sagittal section shows the relation of the mass with the vertebral column. It shows extension up to the ischiorectal fossa and vaginal vault. No e/o erosion of vertebral column or sacrum. **Origin:** Department of Radiology, Civil Hospital, Ahmedabad, Gujarat, India.