ANNALI ITALIANI DI CHIRURGIA

Digital Edition
e-publish on-line
ISSN 2239-253X
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Aggressive angiomyxoma of the pelvis presenting as an obturator hernia

Ann Ital Chir, Digital Edition 2019, 8
pii: S2239253X19031244 - Epub, Nov. 11
free reading: www.annitalchir.com

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Aggressive angiomyxoma of the pelvis presenting as an obturator hernia

INTRODUCTION : *Angiomyxoma is a rare slow-growing soft tissue myxoid cell tumor that usually arises in the pelvis and perineal regions and occurs predominantly in women in the fourth decade. Angiomyxomas usually present as often initially misdiagnosed asymptomatic masses. Most common clinical early diagnoses of aggressive angiomyxomas are in form Of Vulvar Masses, Vulvar Lipomas, Bartholin's Cysts, Levator Hernias, Inguinal Hernias Or Cervical Polyps.*
PATIENTS AND METHODS : *This paper presents the case of the pelvic angiomyxoma diagnosis of a 41 year old with early findings of suspicious obturator hernia during the initial physical examination.*
RESULTS: *The dissection was extended from the right retrorectal area to the ischiorectal cavity and the mass was reached. The capsulated mass of 10*15 cm with soft consistency was completely released and unblocked, it was excised from the abdomen through the incision using wound protection The obturator defect was repaired with interrupted sutures.*
CONCLUSIONS: *Angiomyxoma is a rare, benign and locally aggressive tumor, which can infiltrate locally and present unusually as perineal hernia. Due to its rarity and lack of specific diagnostic requirements, it's difficult to diagnose preoperatively*

KEY WORDS: Angiomyxoma, Obturator hernia, Pelvic mass

Introduction

Aggressive angiomyxoma, which was identified by Steeper and Rosai in 1983, is found most commonly in young female patients (F>M, 6 times more likely). It's a form of neoplasia originated from mesenchyma, most commonly found in the soft tissue of pelvic, perineal and vulvovaginal areas. Clinically, it most often mimics Bartholin's cysts, lipomas, perineal hernias or pedicled soft tissue tumors [1,2].
The optimal course of treatment is the total excision of the mass with tumor free margins. But due to angiomyxoma's infiltrative nature, there's a high rate of local recurrence after total excision.

In this paper we aimed to present this case with early findings of suspicious obturator hernia during the initial physical examination including digital rectal examination. This work is reported in line with the Surgical Case Report Guidelines (SCARE) criteria [3].

Case Presentation

41-year-old female with a pelvic pain and a lump during coughing and straining in the right pararektal area was admitted to the general surgery out patient clinic. The patient had no history of surgery and any systemic diseases. No family history of the patient was found. She was a non-smoker person.
During initial physical examinations, the vitals were stable, general condition was in order, the patient was fully conscious and cooperative. Abdominal examination was normal. Right gluteal area examination was consistent with suspicious lump palpation in paraanal region during coughing. This suspicious lump pushes up to rec-

Pervenuto in Redazione Agosto 2019. Accettato per la pubblicazione Ottobre 2019
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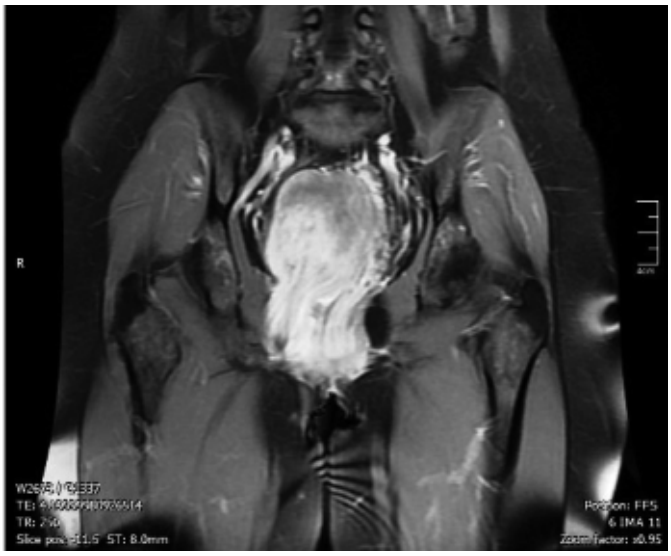

Fig. 1: MRI was showed. A 9*7 cm wide mass legion, which extends from para-rectal through ischiorectal regions was detected.

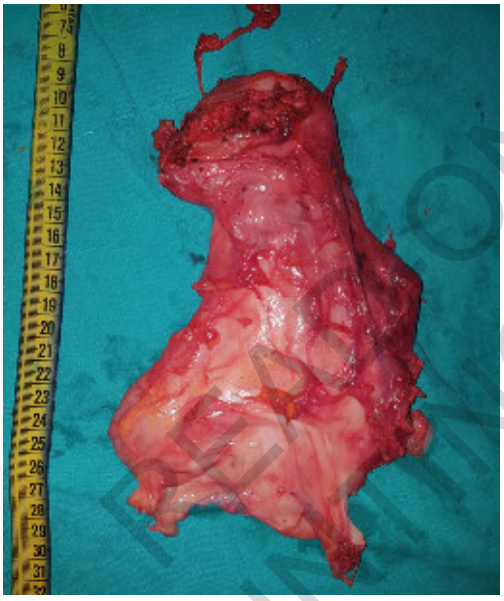

Fig. 2: The specimen had a capsule, 18*10 cm wide, had a soft consistency, was pink-gray colored and shiny.

tum was detected during rectal examination. Howship–Romberg was (+). When an abdominal positive pressure-enhancing reflex is performed on the patient, a pseudomass that is mobile and palpable at the skin level with straining, that could be compatible with a hernia sac was palpated in the perianal region.

A pelvic MRI was performed with a perineal hernia prediagnosis. A 9*7 cm wide mass legion, which extends from para-rectal through ischiorectal regions was detected. The results were inconclusive on the organ the lesion stems from; it has either genital or mesenchymal roots (Fig. 1). Rectosigmoidoscopy findings showed a normal rectal mucosa. Gynecological exam findings showed normal genital organs. The patients' tumor markers and laboratory parameters were in normal ranges.

The operation was performed under general anesthesia in Lloyd Davis Position by a team of colorectal surgeons. The abdomen was insulated with closed technique and a 10mm trocar was placed from cranial 2 cm above the umbilicus for a camera. 2 trocars, one 10mm and one 5mm, were placed from the right paramedian region; one 10mm trocar was placed from the left paramedian region. After routine exploration, pelvic peritoneum was opened and mesorectum was freed with sharp dissection in the caudal plane. The dissection was extended from the right retrorectal area to the ischiorectal cavity and the mass was reached.

The capsulated mass of 10*15 cm with soft consistency was completely released and unblocked, it was excised from the abdomen through the incision using wound protection The obturator defect was repaired with interrupted sutures. One suction drain was placed in Douglas. The patient had no specific post-operative complications and was discharged 5 days after the operation. Upon 22 months of follow-up, there has been no clinical or radiographic (MRI) recurrence and the patient is still attending regular follow-up controls.During histopathological examination, the specimen had a capsule, 18*10 cm wide, had a soft consistency, was pink-gray colored and shiny (Fig. 2).

Immunohistochemical stain results for estrogen and progesterone receptors and Desmin and SMA markers were positive. CD34 MUC4, S100, Beta-catenin results showed negative staining (Fig. 3).

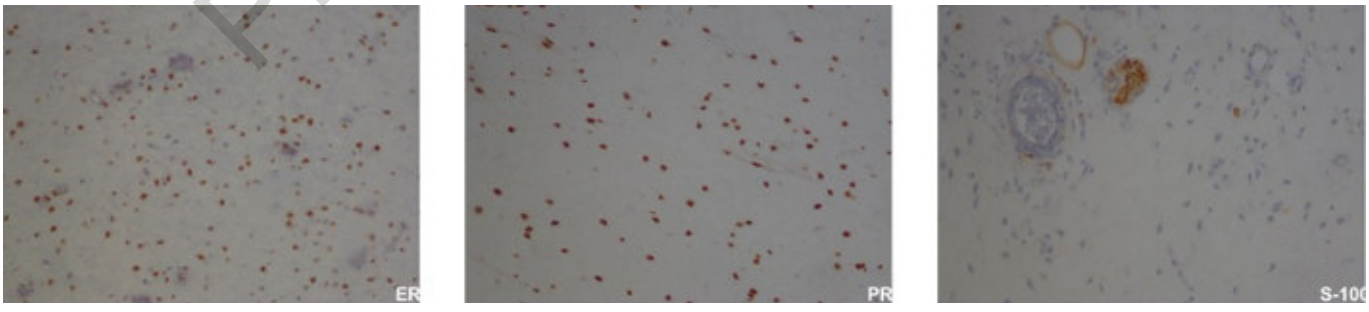

Fig. 3: Immunohistochemically tumor cells showed strong diffuse positivity with estrogen and progesteron receptor but showed negativety with S-100 (x100).

A written informed consent for publication of her clinical details and clinical images was obtained from the patient.

Discussion

Aggressive angiomyxoma is a rare aggressive mesenchymal tumor, which stems from myofibroblasts of pelvis and perineum. It doesn't have a conclusive pathogenesis. Recently, a translocation at chromosome 12 with a consequent aberrant expression of the high mobility group protein isoform I-C (HMGI-C) involved in DNA transcription has been demonstrated [4].

Aggressive angiomyxoma is a morphologically distinctive myxoid neoplasm of soft tissues. The tumor is composed of spindled or stellate cells within a loose matrix with some collagen formation, vessels of varying size and myxoid stroma in a myxoid background. The vessels are prominent and they are thin or thick walled and variable sized. The myxoid background may contain hemorrhage and collagen fibers. The cellularity is generally low and the cells have bland hypercromatic nuclei with centrally located nucleoli. Mitotic figures are rarely seen. This neoplasm tends to locally recur despite its bland morphology. Immunohistochemically Desmin, Smooth Muscle Actin (SMA), Estrogen Receptor (ER) and Progesteron Receptor (PR)are positive; S100 and keratin are negative [5]. Myxoid liposarcomas, sarcoma botrioids, myxoid variant of malignant fibrous histiocytomas, nerve sheath myxomas and secondary myxoid and other soft tissue tumors should be excluded during myxoma differential diagnosis [6].

There are no specific immunohistochemical markers, however most aggressive angiomyxomas stain positive for desmin, smooth muscle actin, muscle-specific actin, vimentin and characteristically for oestrogen and progesterone receptors suggesting a possible hormonal cause for these tumors. It's challenging to diagnose aggressive angiomyxoma during the preoperative period, a definitive diagnosis could be made only after a histopathological examination. Our patient tested positive for estrogen receptors in tumor tissue. Existence of these receptors in the tumor tissue supports the theory of hormonal factors playing a role in the development of these tumors. Most cases of aggressive angiomyxomas usually present as asymptomatic masses often being initially misdiagnosed [7].

Most common clinical symptoms are vulvar masses, vulvar abscesses, lipomas, Bartholin's cysts, Gartner cysts, vaginal cysts, levator hernias, pelvic hernias and cervical polyps [7,8]. Our patient had similar complaints and symptoms to obturator hernia. Obturator hernia is a pelvic hernia due to herniation of obturator nerves and muscles out of the obturator foramen and it makes up 0.05 to 1.4% of all abdominal wall hernias [9].

Radiological imagings play a critical role in sizing the pelvic mass, the choice of surgical approach and detection of any possible recurrence. MRI scans are particularly helpful for the diagnosis. MRI is more specific than CT and it's the preferred method of imaging for pelvic legions [10,11]. Surgery is the recommended treatment and it requires an excision as wide as possible. Complete surgical excision with tumor-free margins is still accepted as the main treatment method for aggressive angiomyxomas [6].

Ineffective operation is the most important factor for recurrence. Tumor cells are highly capable of local infiltration. There were even reports of recurrence in clinical cases with large R0 resection. After an effective and reliable surgical operation, the recurrence rate was reported somewhere between 30-50% [12]. Since the current follow-up period is 22 months, we may think of R0 resection performed because of the disease have not recured yet. Radiotherapy and chemotherapy aren't often considered for primary therapy due to low mitotic activity maybe therefor most studies haven't shown a significant advantage in favor of radiotherapy [8,13] .

Although surgery remains the standard of care, the medical treatment of AAM with a gonadotropin releasing hormone agonist in the primary or adjuvant setting may offer an alternative to radical surgery.

In primary or recurring aggressive angiomyxoma cases GnRH agonist therapy yielded successful results and there has been a significant regression in tumor tissue [14].

Conclusion

Angiomyxoma is a rare, benign and locally aggressive tumor, which can infiltrate locally and present unusually as perineal hernia. Due to its rarity and lack of specific diagnostic requirements, it's difficult to diagnose preoperatively. It shouldn't be overlooked during differential diagnosis of pelvic masses. The principle treatment should be complete surgical excision with tumor-free margins but the patients should be kept on a close watch during remission due to high local recurrence rates.

Riassunto

L'angiomixoma è un raro tumore delle cellule mixoidi dei tessuti molli a crescita lenta che di solito si manifesta nella regione pelvica e perineale, prevalentemente in donne nel quarto decennio di vita. Gli angiomixomi di solito presentano masse asintomatiche spesso inizialmente esposte ad errori di diagnosi. Le diagnosi cliniche iniziali più comuni in caso di angiomixomi aggressivi sono sotto di masse vulvari, lipomi vulvari, cisti di Bartolino, ernie del muscolo levatore, ernie inguinali o polipi cervicali.

Viene qui presentato il caso di un angiomixoma pelvico di un uomo di 41 anni con iniziale sospetto di ernia otturatoria durante l'esame fisico iniziale.

La dissezione chirurgica è stata estesa dall'area retrorettale destra alla cavità ischiorettale e la massa è stata individuata. Si trattava di una massa capsulata di 10 x 15 cm con consistenza morbida, che è stata completamente liberata ed asportata dall'addome attraverso l'incisione facendo attenzione alla protezione della ferita. Il difetto dell'otturatore è stato riparato con suture a punti staccati.
L'angiomixoma è un tumore raro, benigno ma localmente aggressivo, che può infiltrarsi localmente e presentarsi insolitamente come ernia perineale. A causa della sua rarità e della mancanza di requisiti diagnostici specifici, è difficile diagnosticare prima dell'intervento

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