Abstract

Adnexal masses are commonly encountered in gynaecological practice and often present both diagnostic and management challenge. We present a case of vascular leiomyoma in an adolescent female where the presence of a rapidly growing abdominal mass created a diagnostic and management challenge. Only after surgical exploration, histopathology and immunohistochemistry, a final diagnosis of giant vascular leiomyoma (a benign tumor) was made.

Keywords: adnexal, leiomyoma, giant, vascular, adolescent

Rapidly growing giant adnexal mass in an adolescent female- a diagnostic dilemma

Massa anexial com rápido crescimento numa adolescente – um dilema diagnóstico

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BACKGROUND/ INTRODUCTION

Leiomyomas represent the most common gynaecologic & uterine neoplasms. Most of them are detected in women of middle aged group (>30 yrs). These histologically benign tumors, which originate from smooth muscle cells, usually arise in the genitourinary tract but may arise in nearly any anatomic site. Typical appearance of uterine leiomyomas are easily recognized on imaging. However, the ones with unusual growth pattern or in unusual location and at unusual age group form a diagnostic challenge for the clinician. They may mimic malignancies and therefore, result in serious diagnostic errors.

CASE PRESENTATION

A young Asian girl of 17, presented to us with painless lump and distension of abdomen associated with sudden increase in size over a span of four months. There was no history of nausea, vomiting, loss of weight or appetite. There was no history of bowel and bladder irregularity. Menarche was at 13 and no menstrual irregularity was reported. She denied prior sexual activity and use of hormones. Her past medical and surgical history was non contributory. On examination, a pelvic mass of 30 weeks size was palpable. It was firm and non tender with restricted mobility and palpable on per-rectal and as well
as per-vaginal examination. A pelvic sonogram showed a mixed solid cystic mass. Urine HCG test was negative. Repeat ultrasonography revealed a vascular heterogeneous abdominal-pelvic mass measuring 22×20×20 cm with multiple cystic areas arising from right adnexa with a suspicion of ovarian origin. MRI of the abdomen & pelvis showed a right adnexal mass occupying nearly whole of abdominal cavity; the origin was still not clear (Fig.1a,b). Probability of right ovarian mass was suggested. Serum CA125, α fetal protein & Adenosine Deaminase levels were insignificant. The serum levels of β HCG and LDH were also within normal limits.

Gross pathologic examination revealed a large grayish brown mass measuring 25×22×20 cm. The outer surface was shiny, congested with prominent veins & appeared to be covered by a thin capsule which was intact. Cut section was variegated with central yellowish areas having small cysts filled with mucoid material and a peripheral grayish brown solid fleshy area (Fig.3).

Multiple representative sections were submitted for histopathological examination. The microscopy showed a varied morphology (Fig 4). There were areas showing intermediate sized spindle shaped cells with spindled to ovoid nuclei; in bundles and cords with no particular arrangement and surrounded by abundant hyalinization. No atypia/mitoses was seen (Fig.5). Areas showing marked proliferation of small & medium sized blood vessels were seen. These vessels were not interconnected and the endothelial lining did not show any atypia. The sections from yellowish area showed large areas of necrosis which was of hyalinizing type. The capsule was intact, thin and fibrous. A tentative diagnosis of benign spindle cell lesion was made and as leiomyoma is the first differential diagnosis; smooth muscle actin (SMA) Immunohistochemistry was performed which came out to be positive (Fig.6a,b); thus making a final diagnosis of giant vascular leiomyoma undergoing hyaline degeneration.
DISCUSSION

Accurately diagnosing an adnexal mass has become a challenge given the vast diagnostic possibilities. The broad differential diagnosis of an adnexal mass includes lesions of infectious origin, such as a hydrosalpinx or tubo-ovarian abscess caused by pelvic inflammatory disease; physiologic or functional cysts; endometriomas; both benign and malignant neoplasms, and masses originating in organs or tissues proximal to the adnexa. Important considerations in arriving at the most probable diagnosis are age of the patient, clinical history, findings on physical examination and results of radiologic and laboratory studies.

Leiomyoma of the female genital tract is the most common tumor of the female pelvis. Although this tumor is present in approximately one-third of women of reproductive age, the occurrence of leiomyoma in females under 20 years is rare. Also, it has been observed to have a slow growth pattern. In our case the patient was notably only 17 yrs old and presented with a rapidly growing abdominal mass. Ultrasound remains the initial modality of choice but is neither as sensitive nor as specific as MRI. The leiomyomas usually appear on the T2wt images as sharply margined homogenous areas of decreased signal intensity. Degenerated, cystic and fast growing vascular leiomyomas, however show variable MR signal characteristics and this can create a suspicion of ovarian neoplasm particularly in case of a large adnexal mass where it is difficult to assess its origin. Pedunculated lesions exceptionally can have obscure margins and maybe mistaken for a lesion of ovarian origin.

The abovementioned pitfalls of USG & MRI in our case led to a presumptive diagnosis of an ovarian neoplasm. The normal CA-125, LDH, β hCG and α fetoprotein levels negated the possibility of epithelial and germ cell ovarian malignancies respectively, but considering the adolescent age of the patient, the possibility of other benign & non epithelial malignant lesions still had to be ruled out. Only after surgical exploration, the dilemma was cleared as the ovaries were found to be normal and the tumor mass was seen attached to the broad ligament and uterine cornu. Now the main differentials were leiomyoma, leiomyosarcoma, malignant mixed mullerian tumor or rare possibility of a broad ligament fibrothecoma. Microscopy revealed large areas of hyalinization necrosis & intermediate sized plump spindle shaped tumor cells which did not have the characteristic spindle shape with cigar shaped nuclear morphology of a smooth muscle. Absence of nuclear atypia & mitotic figures favored a benign tumor but large areas of necrosis were misleading. It is very important to distinguish coagulative from hyalinizing necrosis as the presence of coagulative necrosis, even in absence of significant atypia would lead to a diagnosis of sarcoma. The marked vascular proliferation also created a diagnostic dilemma, however these vascular channels were not interconnected.
not lined by atypical endothelial cells & were present in focal areas. A positive immunostaining for smooth muscle actin(SMA) finally clinched the diagnosis as giant vascular leiomyoma. It could be a parasitic subserosal uterine or a primary broad ligament leiomyoma. 

A review of the literature revealed a paucity of cases of leiomyomas in adolescents and the majority of cases showed symptoms like abdominal lump and menstrual irregularities. The first reported case of a leiomyoma in an adolescent girl of 13 years was in 1969 by Wisot et al, and they performed myomectomy because of profuse bleeding and anemia\textsuperscript{10}. Our case demonstrates the diagnostic challenge posed by the atypical clinical, imaging & histological features of a very common uterine tumor, in terms of its presentation, appearance, location and its mimicry of a primary ovarian tumor in an adolescent female.

REFERENCES


