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Case report

Aggressive angiomyxoma presenting as a huge intra-abdominal mass in a pregnant Nigerian woman: a case report

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Abstract

Aggressive angiomyxoma is a rare, locally aggressive myxoid mesenchymal tumour that predominantly arises from the pelvis and perineum of young adult females. Occasional cases have been reported at unusual sites. Oestrogen and progesterone may play a role in the development of the tumour especially during pregnancy. A 25 year old Nigerian woman presented with severe abdominal pain shortly after a fresh still birth. Clinical and radiological evaluation revealed a massive intra-abdominal mass which was excised and pathologically diagnosed as aggressive angiomyxoma. The follow up period has been uneventful. This case demonstrates that aggressive angiomyxoma may present as a primary intra-abdominal mass and thus should be considered in the differential diagnosis of such lesions as this will greatly impact on its surgical management. Moreover, pregnancy appears to influence rapid tumour growth and may result in massive tumour sizes. Long term patient follow-up is routinely advocated for the early detection of tumour recurrence.

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Introduction

Aggressive angiomyxoma (AA) is a rare, indolent, slow growing, locally aggressive myxoid mesenchymal tumour that preferentially arises from the connective tissues of the pelvis and perineum of young adult females although occasional cases have been reported at unusual sites [1,2]. Since it was first described in 1983 fewer than 350 cases have been reported in the literature [3]. It is often mistaken for other more common lesions since pre-operative diagnosis and assessment of deep tissue involvement are often difficult [4]. Thus it is typically associated with a high rate of local recurrence although rare cases of metastasis typically involving the lungs have been reported in the literature [5,6]. AA's which are typically large and lobulated have been associated with characteristic light microscopic, immunohistochemical and molecular genetics features [3,7]. It has been suggested that oestrogen and progesterone may play a role in the development of the tumour since it predominantly affects premenopausal women and typically expresses receptors for these hormones [1,3]. Here we describe the case of an intra-abdominal AA in a young pregnant Nigerian woman. To the best of our knowledge, this may be the first report of such a case in Nigeria.

Patient and observation

A 25 year old Para 2+0 (none alive) seamstress presented at the emergency unit of our hospital having been referred from a private hospital on account of severe abdominal pain following a fresh still birth with an ultrasound diagnosis of ovarian mass. The labour was uneventful. However, she suddenly developed colicky left sided abdominal pain about 5 hours after the delivery which was severe enough to disturb her activities. It later became generalized with no known aggravating or relieving factors. It was associated with dizziness but no fainting attack. She claimed she had always felt she had an intra-abdominal mass long before the index pregnancy without any obvious abdominal swelling. The antenatal period was uneventful while the previous pregnancy also resulted in a fresh still birth. Examination revealed a young woman in painful distress. She was restless and mildly pale but not jaundiced or dehydrated. The respiratory rate was 28 cycles per minute although there was good air entry into both lungs. She had a pulse rate of 102 beats per minute while the blood pressure was 104/72 mmHg. A 20cm X 30cm mass was palpable in the left lower abdominal quadrant. The mass was mobile and moderately tender. The liver, spleen and kidneys were not palpable. The uterus was 18 weeks in size and well contracted. There was no ascites. A diagnosis of torsion of an ovarian mass was made and she was immediately prepared for an exploratory laparotomy under general anaesthesia. The full blood count, electrolyte and urea, and urinalysis were all within normal limits. At surgery we found a haemorrhagic mass firmly attached to the mesentery with small bowel loops adherent to it. There was about 300mls of haemorrhagic peritoneal fluid. The liver, spleen, uterus, ovaries and fallopian tubes were essentially normal. The mass was completely excised along with the adherent bowel loops and an end to end anastomosis of the small bowel loops was done. The postoperative period was uneventful and the patient was discharged on the seventh. The gross pathology was that of a soft, fleshy grayish-white mass measuring 21.0 X 20.0 X 14cm and weighing about 22kg with (**Figure 1**, **Figure 2**). Loops of the ileum were loosely attached to the mass although there was no evidence of being invaded by the tumour. Microscopic examination showed a tumour composed of abundant myxoid stroma containing numerous blood vessels of varying calibers (**Figure 3**). Perivascular collagen condensation was noted (**Figure 4**). The tumour cells were scanty and spindle-to-stellate shaped (**Figure 5**). The tumour was seen extending into the muscle wall of the attached ileum (**Figure 6**). The stromal cells were positive for vimentin, desmin as well as estrogen and progesterone receptors. A pathological diagnosis of aggressive angiomyxoma was made. The patient was, therefore, placed on long term clinical and radiologic follow up which has been uneventful.

Discussion

Although majority of aggressive angiomyxomas are known to originate from within the pelvis and perineal region in women, occasional cases have also been reported outside these anatomic sites. The buttocks, retroperitoneum and inguinal regions have been implicated in such cases [2,3]. The unusual location of the tumour in this index case shows that aggressive angiomyxomas may also arise from within the intra-abdominal cavity, particularly the mesentery. At initial clinical evaluation aggressive angiomyxoma is often mistaken for other more common lesions [8]. The diagnostic confusion characteristically encountered in the clinical assessment of this tumour is further magnified when it is encountered at unusual body sites as in this case. Irrespective of the anatomic site of origin,

immunohistochemal and ultrastructural studies seem to indicate that aggressive angiomyxoma probably arises from the primitive mesenchymal cells that are located in these sites and are capable of fibroblastic and myofibroblastic differentiation [9]. In fact the tumour cells are known to show chromosomal translocations involving 12q13-15 which have been reported in a variety of other mesenchymal neoplasms [3,7]. These translocations affect the HMGA2 gene, a transcription factor that functions during embryonic life and not typically seen in adult tissues. Immunohistochemical identification of the gene product may therefore, play important diagnostic roles in the future [3,7]. Aggressive angiomyxomas tend to displace adjacent organs without actually invading them. Nevertheless, in longstanding cases, the tumours eventually invade adjacent organs as seen in our patient in whom the tumour had already started infiltrating the wall of the ileum [2]. With time they may eventually grow into a huge tumour which may completely displace the affected abdominal or pelvic organs [2]. The large tumour size in this index case may, however, have resulted from a rapid growth of the tumour under the influence of oestrogen and progesterone which are produced in large amounts during pregnancy. These hormones may actually influence the growth of the tumour since AA predominantly affects premenopausal women and expresses both oestrogen and progesterone receptors [1,3]. Moreover the tumour is known to respond to hormonal manipulations using gonadotropin releasing hormone (GnRH) analogues [8]. In fact, several reports have shown complete radiographic tumour regression [3]. These may offer an alternative to radical surgery in the primary or adjuvant setting.

As shown in this case, the pre-operative diagnosis of aggressive angiomyxomas is difficult and typically made only at histology. This is because the tumour is extremely rare and not associated with any characteristic clinical features. This factor is known to contribute significantly to the high rate of local recurrence associated with this neoplasm as the extent of the tumour is usually not apparent at surgery thus leading to incomplete tumour resection [3]. Nevertheless, early recognition is critical and requires a high index of suspicion. To this end, a few studies have shown that aggressive angiomyxomas have characteristic appearances on CT and MR imaging techniques with clear delineation of the extent of the tumours, especially in relation to the pelvic diaphragm which is critical to determining the best surgical approach [4,10]. The tumour shows attenuation less than that of muscle on CT scan while they show high signal intensity on T2-weighted MRI [11]. Aggressive angiomyxoma must be differentiated from the more common benign and malignant myxoid tumours. The distinctive vascular component (**Figure 3**), perivascular rings of condensed collagen (**Figure 6**) and soft tissue infiltration characterized by entrapment of muscle, nerve and adipose tissue (**Figure 4**) may be useful in this regard [9]. There are no specific diagnostic immunohistochemical markers but the tumours cells typically show strong positivity for vimentin, desmin, smooth muscle actin, muscle specific actin as well as oestrogen and progesterone receptors while the reaction for CD34 is variable [1,3,11]. Surgery, with wide local excision, remains the mainstay of treatment although several other modalities including radiotherapy and chemotherapy have been tried with unsatisfactory results [8,9]. Complete tumour resection is, however, a daunting task owing to the infiltrative nature of the tumours and the paucity of a pre-operative diagnosis. Therefore, long term follow-up with imaging studies is typically recommended. Tumour recurrence (32%-72%) may be as early as nine months or as long as 20 years after surgery [2].

Conclusion

Aggressive angiomyxoma may present as a primary intra-abdominal mass and thus should be considered in the differential diagnosis of such lesions as this will greatly impact on its surgical management. Moreover, pregnancy appears to influence rapid tumour growth and may result in massive tumour sizes. Long term patient follow-up is routinely advocated for the early detection of tumour recurrence.

Competing interests

The authors declare no competing interest.

Authors' contributions

Donatus Sabageh: Wrote the first and final drafts of the manuscript. He was the pathologist in charge of the case. He also took the gross pictures and micrographs. **Adewale S. Adeyemi:** was the obstetrician in charge of the case. He reviewed the first and final drafts of the manuscript for important intellectual content. **Adetunji S. Oguntola:** was the surgeon in charge of the case, he initiated the write-up. He reviewed the first and final drafts

of the manuscript for important intellectual content. All authors have read and agreed to the final version of this manuscript and have equally contributed to its content and to the management of the case.

Figures

Figure 1: The tumour which is well circumscribed but unencapsulated is seen here with the attached loops of small bowel

Figure 2: Cut surface of the tumour shows a soft fleshy tumour with entrapped loops of small bowel

Figure 3: The tumour is composed of varying calibers of blood vessels within a myxomatous stroma (X100, H&E)

Figure 4: Condensed perivascular collagen rings typical of aggressive angiomyxoma (X400, H&E)

Figure 5: High power view showing individual spindle and stellate shaped tumour cells within a myxoid stoma (X300, H&E)

Figure 6: The tumour can be seen here extending into the serosa and muscularis propria of the ileum resulting in the splaying of the muscle fibres (X100, H&E)

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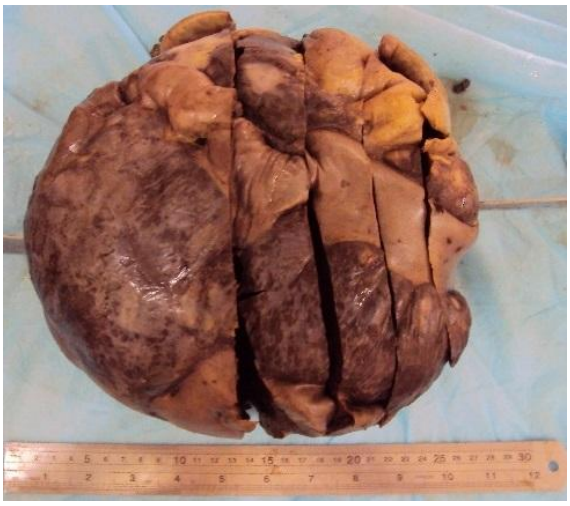

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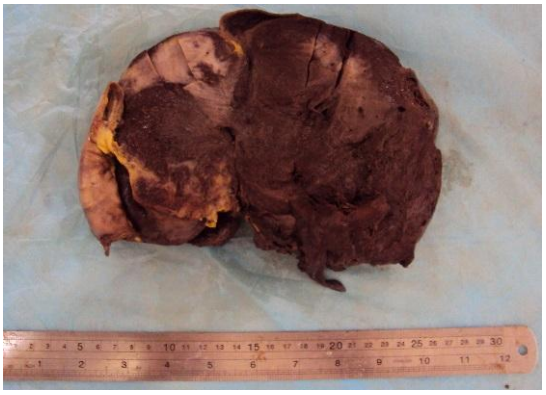

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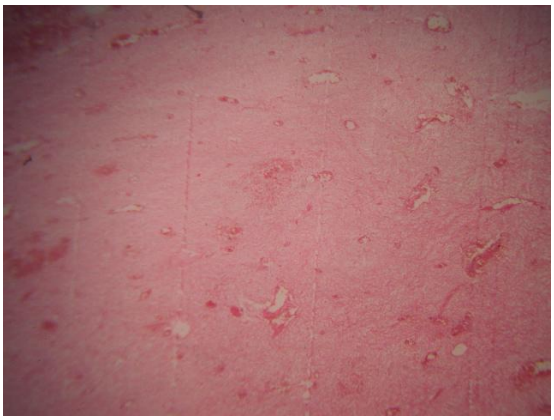

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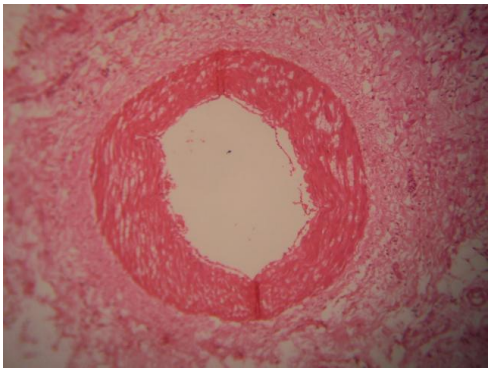
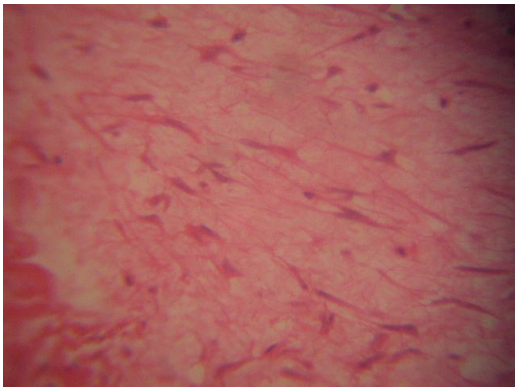


Figure 4: Condensed perivascular collagen rings typical of aggressive angiomyxoma (X400, H&E)



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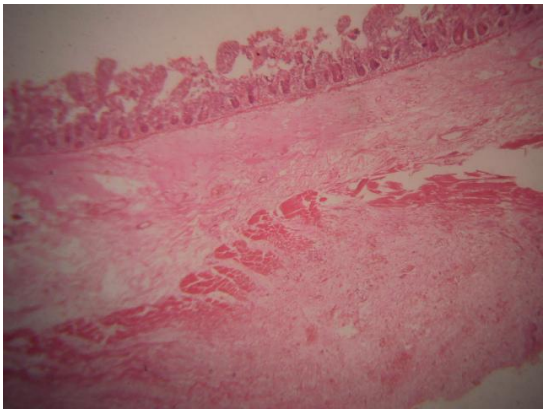

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