

Atypical Leiomyoma with Amianthoid-like Fibers (ALWAF), causing Rupture Uterus, Haemoperitoneum and Mortality- A Case Report

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ABSTRACT

Atypical Leiomyoma with Amianthoid-like Fibers (ALWAF) is a very rare entity with a couple of cases in the literature. Amianthoid fibers are thick acellular mats composed of crystalline collagen fibers. This is an unusual case of a 53-year-old female with Chronic Kidney Disease (CKD), who presented with pain in the abdomen for three weeks. Computed Tomography (CT) showed a degenerated subserosal leiomyoma in the posterior wall of the uterus which had ruptured leading to haemoperitoneum. An emergency exploratory laparotomy with simple hysterectomy was done. Histopathology revealed a ruptured atypical leiomyoma with abundant amianthoid-like fibers. The patient expired seven days after surgery due to hypovolaemic shock. Rupture of the uterus in leiomyoma with amianthoid fibers has not been reported in the literature. Awareness of this entity is important in view of acute catastrophic consequences like haemoperitoneum, hypovolaemic shock, and death.

Keywords: Crystalline collagen fibers, Degenerative changes of leiomyomas, Hypovolaemic shock

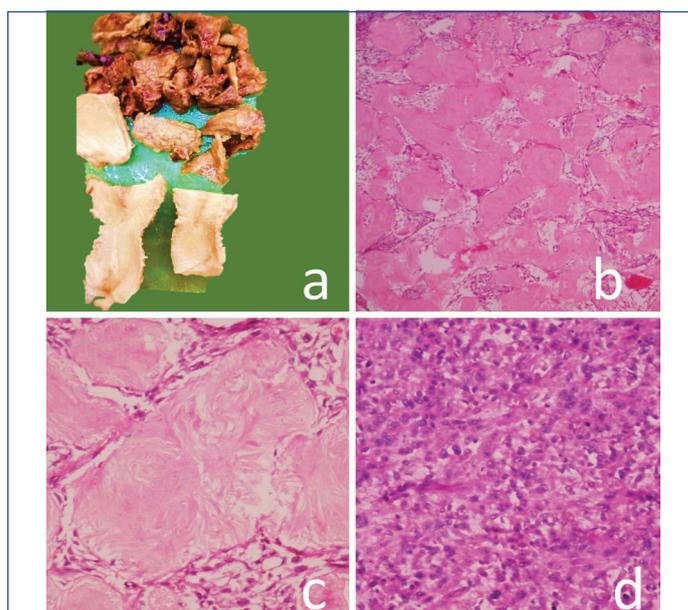
CASE REPORT

A 53-year-old female, a known case of Chronic Kidney Disease (CKD) with hypertension on dialysis, presented with acute pain in abdomen to the emergency services. She had a history of abdominal pain for three weeks and was referred to the tertiary care centre for increasing pain and worsening of general condition. Abdominal distension, tenderness, and guarding were present. Her blood pressure was 140/90 mmHg. She was evaluated in the General Surgery and Gynaecology Departments. On Contrast Enhanced Computed Tomography (CECT) of the abdomen and pelvis, a defect in the posterior wall of the uterus with lobulated periuterine haematoma with haemoperitoneum, probably secondary to degenerated subserosal fibroid was found. An emergency exploratory laparotomy with hysterectomy was done. A clinical diagnosis of endometrial adenocarcinoma leading to rupture uterus or ruptured ovarian tumour were made and at laparoscopy, the fundus of the uterus had a gun-shot appearance.

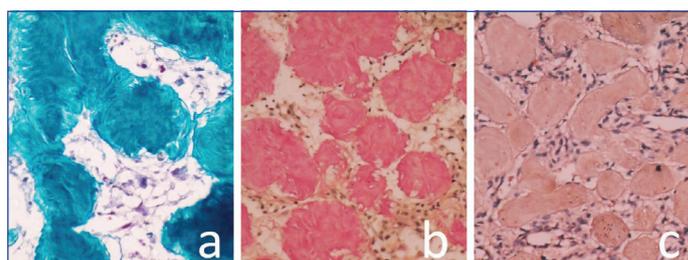
A simple hysterectomy specimen along with multiple brown bits of uterine tissue was received [Table/Fig-1a]. The uterine fundus showed a rupture of 3×2 cm with ragged haemorrhagic margins on its posterior aspect. On microscopy, a spindle cell tumour was identified within the myometrium, which showed a classic picture of an atypical leiomyoma with pleomorphic spindle cells arranged in fascicles. Large mats of acellular amianthoid-like fibers were seen amidst the atypical spindle cells [Table/Fig-1b,c]. On higher magnification the spindle cells showed plump pleomorphic nuclei with moderate cytological atypia [Table/Fig-1d]. No mitosis or necrosis were seen. On special stains, amianthoid-like fibers showed sea green colour on Masson Trichrome [Table/Fig-2a] and bright pink colour on Elastic Van Gieson stain [Table/Fig-2b], while Congo red [Table/Fig-2c] was negative. Desmin [Table/Fig-3a] and Smooth Muscle Actin (SMA) [Table/Fig-3b] were positive, while DOG1, CD10, CD34, and Cyclin D1 were negative. MIB1 was 0. A diagnosis of Atypical Leiomyoma with Amianthoid-like Fibers (ALWAF), leading to uterine rupture was rendered. The patient expired seven days after surgery due to hypovolaemic shock.

DISCUSSION

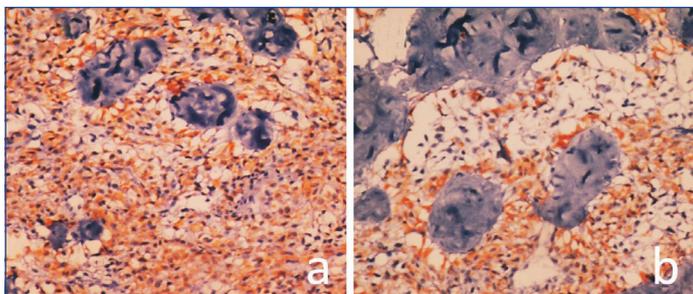
The ALWAF is an extremely rare entity with a couple of cases reported in the literature. Amianthoid-like fibers are thick, acellular mats of



[Table/Fig-1]: a) Hysterectomy specimen: Uterus measured 8×4×2 cm in size with cervix measuring 4 cm in length. Multiple brownish bits of uterine tissue were also received, aggregating to 11×8×2 cm; b) Large mats of acellular eosinophilic fibrillary amianthoid-like fibers surrounded by an atypical spindle cell proliferation (H&Ex400); c) Mats of amianthoid-like fibers showing characteristic fibrillary pattern (H&Ex400); d) Atypical leiomyoma composed of pleomorphic spindle cells showing moderate cytological atypia (H&Ex400).



[Table/Fig-2]: a) Masson trichrome showed mats of amianthoid-like fibers with sea green colour (x400); b) Elastic Van Gieson stain showed mats of amianthoid-like fibers with rose pink colour (x400); c) Congo red showed congophilic nature of the fibers but no apple green birefringence on polarisation was seen (x400).



[Table/Fig-3]: Immunohistochemistry; a) Desmin showed strong positivity in atypical spindle cells (x400); b) Smooth Muscle Actin (SMA) showed strong positivity in atypical spindle cells (x400).

collagen, composed of crystalline fibers, surrounded by a spindle cell proliferation.

Uterine leiomyomas are benign mesenchymal tumours of smooth muscle origin. About 90% of leiomyomas are of the conventional type while 10% show a spectrum of morphological types. A variety of histological subtypes like cellular leiomyoma, atypical leiomyoma, mitotically-active leiomyoma, lipoleiomyoma, epithelioid leiomyoma, angioleiomyoma, and cotyledonoid type have been described [1,2]. Atypical leiomyomas are smooth muscle tumours with either focal or diffuse, moderate to severe cytological atypia, without coagulative necrosis and with a very low mitotic index [2].

Leiomyomas with amianthoid fibers or amianthoid-like fibers are very unusual leiomyomas, and their incidence is restricted to a couple of case reports in the literature. Spontaneous rupture of the uterus, however, has not been reported, and to the best of our knowledge, this is the first case in the literature describing spontaneous rupture leading to a haemoperitoneum, hypovolaemic shock, and death.

Collagen can be classified into four types I, II, III and IV based on fibrillar and non fibrillary organisation, molecular and immunohistochemical differences. Type I, II and III are crystalline and type IV is amorphous appearing [3]. The term amianthoid was used for degenerating costal cartilage for macroscopically and microscopically observed fibers [4]. Amianthoid fibers are acellular, eosinophilic, extracellular mats composed of crystalline collagen fibers. Immunohistochemistry shows positivity for collagen type I at the centre and collagen type III at the periphery [5]. Amianthoid fibers vary from other collagen fibers in thickness, contour and the arrangement of collagen. They have a width of 1000 nm and a normal banding of 52-62 nm, and are 10 times the thickness of normal collagen [4]. Normal or native collagen has a thickness of 35-120 nm [6]. The term amianthoid-like fibers is used if the thickness of the fibers does not fall into the specified range of width 1000 nm and thickness more than 200 nm of amianthoid fibers [7]. Ultrastructural studies are required to confirm the diameter of the fibrils and hence the fibrils were termed as "amianthoid-like", as the authors were unable to perform ultrastructural studies.

The exact pathogenesis of amianthoid fibers is unclear. Various hypotheses have been put forth which include the falling together of pre-existing collagen fibrils or unmasking of collagen from the interfibrillar matrix [4]. Anoxia has been implicated in the degeneration

of perivascular collagen [8]. Many also believe that the amianthoid fibers, in soft tissue tumours, results from active secretion and deposition by the tumour cells, and thus is degenerative in nature, leading to the rupture of the uterus and death of the index patient. In a case of chondrosarcoma described by Ghadially FN et al., amianthoid fibers found were 1000 nm thick and were located in the matrix of the chondrosarcoma. They favoured a degenerative origin for them [6]. Amianthoid fibers are also seen as retrogressive changes that takes place in hyaline cartilage e.g., costal cartilage [4]. Collagen becomes more crystalline with age, through cross-linking and the fibrils become thicker [4]. Exaggeration of this process, with straightening and close-packing of the fibrils results in the formation of amianthoid fibers [4].

Amianthoid fibers are seen in a variety of neoplastic and non neoplastic conditions. They have been often described in intranodal palisaded myofibroblastoma [5]. Case reports of leiomyomas, soft tissue tumours, myofibroblastoma, chondrosarcoma and even meningioma with amianthoid fibers are described in the literature [5,9-11]. Amianthoid fibers have been described in both conventional as well as atypical leiomyomas. However, no case of leiomyosarcoma with amianthoid fibers has been described till date.

CONCLUSION(S)

Rupture of the uterus in leiomyoma with amianthoid fibers is extremely unusual and has not been reported in the literature. This case of rupture of uterus, leading to death as a consequence of the presence of amianthoid-like fibers in a leiomyoma, and awareness of this entity should be promoted because such catastrophic complications can result in death. This case report also expands the readers knowledge of the morphological spectrum of leiomyomas.

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