

Case Report

Giant leiomyoma uterus with myomatous erythrocytosis syndrome: A rare case report

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Abstract

Although leiomyoma uteri are common, giant myomas are rare. The rare occurrence of erythrocytosis, myomatous uterus and persistent restoration of normal haematological values after hysterectomy is known as the myomatous erythrocytosis syndrome. A 46-year-old woman presented with progressive swelling of the abdomen by a hugely protuberant mass for three years. Clinical suspicion was of malignant ovarian tumour. Ultrasonography was inconclusive and CT revealed a huge lobulated heterogeneous soft tissue mass with intralesional vascularity. Her haemoglobin was 22.4 gr/dL and hematocrit of 56%. She underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. On tenth postoperative day her repeat haemoglobin and hematocrit was 19.5 gr/dL and 51.9% respectively. Follow up at six weeks and 6 months after surgery her haemoglobin was 16.5 gr/dL and 42.8% respectively and her hematocrit had regressed to 47.6% and 42.8% respectively. Giant leiomyomas can present an unusual challenge in the management.

Key Words:

Leiomyoma, giant, erythrocytosis, myoma, uterus

Introduction

Leiomyoma are the commonest of all pelvic tumors, observed in 20% women of reproductive age group [1]. Although they are the commonest tumor, giant myomas (11.4 kg or more) are very rare [2]. Myomatous erythrocytosis syndrome is a rare entity [3] and the first case of myomatous erythrocytosis syndrome was reported in 1953 by Thomson and Marson [4]. Less than 40 cases of myomatous erythrocytosis syndrome have been reported in the literature since then [5]. We present a case of myomatous erythrocytosis syndrome in a postmenopausal women presented with giant leiomyoma uterus.

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Case Presentation

A 46-year-old woman presented with progressive swelling of the abdomen, vague abdominal discomfort, lower abdominal and pelvic pain and frequency of urination from last 3 years. The swelling gradually increased and there was off and on breathing difficulties and swelling of bilateral legs from last 6 months. Menstrual history revealed she attained menopause one year back and there was history of heavy flow for one and half years priors to she attained menopause. She had 3 children and her last child birth was 23 years back. There was no significant medical or surgical history. On general examination her vitals were normal; there was bilateral pedal oedema but no lymphadenopathy. On abdominal examination her abdomen was circumferentially distended (Figure1) by a huge abdominal and pelvic mass, which was non-tender, soft elastic in consistency and dull on percussion. On bimanual examination fornices were full and a very large, soft elastic, central mass that filled the entire pelvis and ab-

domen was felt. It was difficult to specify the origin of mass. Her haemoglobin was 22.4 gr/dL and hematocrit of 56%. All other laboratory investigations were within normal limit, including serum CA-125 and papsmear. Abdominal ultrasonography showed a huge heterogeneous soft tissue mass arising from pelvis and occupying almost entire abdomen, uterus was poorly visualized. Contrast CT scan of the abdomen and pelvis revealed a huge lobulated heterogeneous soft tissue mass lesion noted in the pelvic cavity extending into the abdominal cavity. Multiple intralésional vascularity was noted (Figure 2). Sigmoid colon was adherent to the mass. The mass appeared to be originating from uterus.

Figure 1.



Presentation with huge enlarged abdomen

The patient underwent laparotomy with midline vertical incision. At laparotomy a huge multilobbed, semi solid tumour arising from the uterus, with multiple big subserosal fibroids, that filled the entire abdomen was noted that compressed the abdominal organs. Total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed. The tumour mass weighed 11.6 kg and measured 43cm x 32cm x 23 cm (Figure 3). Her intraoperative and postoperative period was uneventful.

On tenth postoperative day her repeat haemoglobin and hematocrit was 19.5 gr/dL and 51.9% respectively. Follow up visit at six weeks and 6 months her haemoglobin was 16.5 gr/dL and 42.8% respectively and her hematocrit had regressed to 47.6% and 42.8% respectively. The patient was doing absolutely fine.

Figure 2.



Contrast CT in axial section showing a large lobulated enhancing mass filling the abdominal cavity along with evidence of intense internal vascularity

Discussion

The largest uterine fibroid ever reported in the literature weighed 63.3 kg; it was removed postmortem in 1888 [6]. A 60.7 kg myoma was removed from a patient in 1930, but she died of pneumonia 48 hours later [2]. The largest uterine tumor ever removed from a patient who survived the procedure weighed 45.4 kg [7] Giant leiomyoma represents a great diagnostic and therapeutic challenge even for the most experienced gynaecologist. Imaging studies (ultrasound, CT, MRI) and tumor markers are helpful to define the extent of the mass and the likelihood of malignancy, respectively. Treatment of leiomyoma should be individualized, depends on symptom severity and patients desire to preserve fertility [8].

The combination of erythrocytosis, myomatous uterus and persistent restoration of normal haematological values after hysterectomy is known as the myomatous erythrocytosis syndrome [3]. Myomatous erythrocytosis syndrome is a very infrequent occurrence in leiomyoma uterus.

Figure 3.



Postoperative specimen- uterus (small arrow) with multiple fibroids and vague worm-like appearance (big arrow) weighed 11.6 kg and measured 43cm x 32cm x 23 cm

In this patient all three diagnostic criteria of the myomatous erythrocytosis syndrome were met. The exact pathophysiology of this condition is unclear. A number of theories have been proposed [5] intramyomatous arteriovenous shunting, substantial myoma size, myoma site, impaired pulmonary function, local hypoxia, change in red cell life span, altered renal erythropoietin (Epo) output, and direct production of Epo by the myomas. Erythropoietin is primarily produced by cells of the renal cortex and stimulates growth and differentiation of the erythrocyte progenitor cells. Both erythropoietin and erythropoietin receptor (Epo-R) are expressed on a great variety of tissues including the uterus. Recent evidence confirmed Epo production by myoma tissues of patients with associated erythrocytosis [9, 10]. It has been hypothesized that erythropoietin may contribute to myoma growth by stimulating angiogenesis. The production of Epo and Epo-R by uterine leiomyoma tissue has been confirmed by immunohistochemical staining and it was speculated that Epo effectively acted on the unusual size of myoma [9]. In

the myomatous erythrocytosis syndrome the estrogen-induced erythropoietin production may be further augmented by local hypoxia due to the rapid growing myomatous tissue, or by paracrine and/or autocrine mechanism [9, 10] Rothman and Rennard [11] suggested that the myoma may compress the renal vessels and thus impair the circulation to the kidney. This might have an effect on the kidney erythropoietin production similar to that found in non-neoplastic renal disease. Such a possibility might seriously be considered in our case since it is consistent with the fact that the polycythaemia has been found usually in association with large myomas. A large abdominal tumour might interfere with pulmonary ventilation, causing hypoxia and consequent stimulation of erythropoiesis by the mechanism described above [11]. This may be in our case since patient presented with large tumour which might cause her respiratory embarrassment. In our case patient had a history of severe menorrhagia and, in spite of that, her haemoglobin was 22.4 gr/dL on admission, erythropoietin induced polycythaemia might probably the cause for this. Recognition of the condition pre-operatively, might be useful-as some authors [11,12] have done-to bleed the patient pre-operatively, store the blood and administer it again during or after the operation should bleeding become excessive. However, to give whole blood during or immediately after the operation in such patient might be dangerous. If blood volume has to be restored, plasma transfusion is the method of choice [12]. In view of the high incidence of uterine myoma and the consecutive expression of erythropoietin and/or Epo-R in normal and myomatous endometrium, it is amazing that the myomatous erythrocytosis syndrome occurs so infrequently. Giant leiomyomas are rare and present an unusual challenge even for the most experienced gynecologist. The rare occurrence of erythrocytosis, myomatous uterus and persistent restoration of normal hematological values after hysterectomy is known as the myomatous erythrocytosis syndrome. Various theories regarding the pathogenesis have been put forward. After hysterectomy, the condition usually disappears.

Acknowledgement

None

Declaration of Interest

None

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