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# THE MYOMATOUS ERYTHROCYTOSIS SYNDROME;

## A CASE REPORT AND REVIEW OF LITERATURE

**Dr. Ayesha Ahmed[1], Dr. Nasira S Dawood[2], Dr. Faiza Ibrar[3]**

1. OB/GYN Resident
   Fauji Foundation Hospital,
   Rawalpindi, Pakistan
2. Professor and Head of Unit-I
   Obstetrics and Gynaecology
   Fauji Foundation Hospital,
   Rawalpindi, Pakistan
   Assistant Professor Obstetrics and Gynaecology
   Fauji Foundation Hospital,
   Rawalpindi, Pakistan

**Correspondence Address:**
Dr. Ayesha Ahmed
H No. 78, Lane 5, Askari 10,
Airport Road, Rawalpindi
ayeshaahmed89@hotmail.com

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**ABSTRACT...** Uterine leiomyomas are the most common benign pelvic tumours in females. Rarely they are associated with symptoms of ectopic hormone production. One of these syndromes is the myomatous erythrocytosis syndrome, which is a triad of polycythaemia, fibroid(s) and resolution of the polycythaemia after hysterectomy. The case presented is of a post-menopausal lady found to have a Hb of 21 during workup of a pelvic mass. A diagnosis of secondary polycythemia due to ectopic erythropoietin production by the fibroid was made. After five sessions of phlebotomiesa target Hb of 15 was obtained for surgery. The haematological values restored to normal three days post-operatively. It has been postulated that all fibroids produce erythropoietin. Recognition and proper management of this condition is important to differentiate between primary and secondary polycythaemia as primary polycythemia is a thromboembolic condition, to avoid un-necessary investigations and to prevent any complications secondary polycythaemia may have per-operatively.

**Key words:** Fibroid, Myoma, Polycythaemia, Erythrocytosis, Gynaecology, Ectopic hormone.

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## NOVELTY

The article describes a rare and interesting case of a post-menopausal lady presenting with a massive fibroid associated with ectopic hormone production leading to polycythemia. To the best of our knowledge this is the first case of its type to be reported in Pakistan and one of the largest fibroids yet reported internationally. We believe our findings would appeal to the readers of this journal as fibroids are the most common pelvic tumors worldwide and recognizing the syndrome along with timely and appropriate management is crucial.

Recognizing the importance of awareness about this syndrome, this article was chosen for presentation at the “Abu Dhabi Obstetrics, Gynecology and Midwifery Congress 2013”.

## INTRODUCTION

Uterine leiomyomas are the most common benign pelvic tumors in females. Usually they cause menorrhagia and pressure symptoms. Rarely they may be associated with symptoms of ectopic hormone production. One of these conditions is the myomatous erythrocytosis syndrome, which is defined by the triad of erythrocytosis, a myomatous uterus and the restoration of normal hematological values after hysterectomy. The rarity with which secondary polycythaemia occurs in cases of uterine fibroids (0.02-0.5%)[1] is interesting considering fibroids are found in upto 77% of histological specimens at hysterectomy. Recognition and proper management of this condition is important to differentiate between primary and secondary polycythaemia, to avoid un-necessary and expensive investigations, to ensure strict monitoring of the patient for thromboembolic events while admitted, to avoid any complications secondary polycythaemia may have per-operatively and finally to confirm the diagnosis after surgery.

## CASE REPORT

The patient, a 70 year old female, post-menopausal for 8 years, presented with the complaint of a large mass in the abdomen for 2 years. The mass was arising from the pelvis and extending towards

the epigastrium. It enlarged slowly over time, was associated with diffuse, dull pain, radiating to the legs and perineum. No history of weight loss, loss of appetite, fever or night sweats. On examination vital signs were normal. Abdominal exam revealed a 36 weeks size mass, solid, mobile, non- tender with irregular contours having a central sulcus. Labs showed a haemoglobin (Hb) of 21.9g/dl, Hct 67,MCV 79, MCH 25 and a MCHC of 32. RBC count was 8.41, all other baseline normal. Blood film showed erythrocytosis. Tumor markers (AFP, CA-125, beta-HCG) were within normal range. ECG had Q waves in anterior wall, and a mild left axis shift noted. On CT scan [Figure-1] a homogenous mass was found to be originating from the pelvis and extending to the epigastrium, measuring 31.2x20.7x35.8cm. Multiple calcific foci were seen within it. Enhancing vessels seen coursing through the mass. Interface with adjacent viscera was intact. The mass was compressing the urinary bladder anteriorly and had pushed the gut loops to the right.

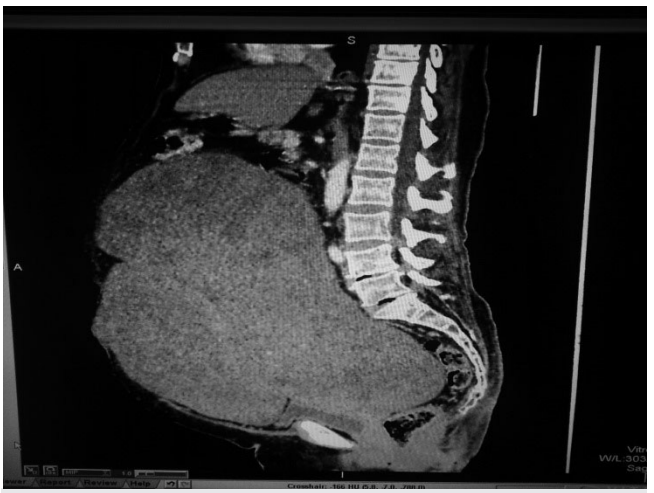

**Figure-1. Sagittal view of CT scan of the patient showing the massive fibroid.**

In view of the polycythemia a haematologist opinion was sought, who advised phelebotomy on alternate days till an appropriate Hb was achieved for surgery. Phelebotomy was done 5 times till a target Hb of 15 and a Hct of 45 was achieved, and then laparotomy was performed. The mass was seen to be a broad ligament fibroid (left side), measuring about 36 weeks in size, having tortuous vessels on the surface, multiple small masses incorporated posterior to the main mass inside the broad ligament. Uterus was hidden behind the mass, 8 weeks in size and rotated on itself, cervix was completely shifted towards left. Round ligaments were identified which were broadened and stretched. Left fallopian tube stretched over the fibroid. Right tube normal. Tissue planes were identifiable. Bladder was stretched over the mass. Left ureter was pushed laterally. A total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed. Post –operatively the patient remained stable. First day post – op, the Hb was 11g/dl. Three days post-op Hb was 10.3g/dl and Hct 32. Long-term Hb measurements remained within the normal range, thus confirming that the fall in Hb was due to reversal of the syndrome and not due to surgical blood loss (post-surgical anaemia). The restoration of haematological values to normal post-operatively fulfils the triad of the myomatous-erythrocytosis syndrome.

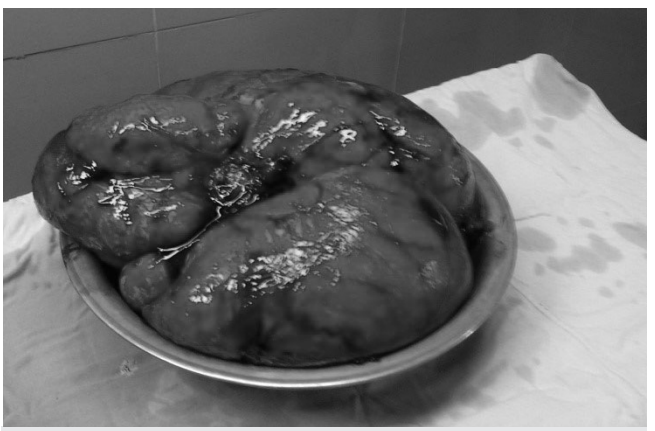

**Figure-II. The fibroid once removed surgically**

## DISCUSSION

The first case of polycythaemia associated with fibroids was reported in 1953. Multiple etiological factors have been proposed since then to explain the development of polycythaemia. These include erythropoietin (EPO) or erythropoietin-like substance production by the leiomyoma itself. Another theory is that tissue hypoxemia in the myomata, intra-uterine shunting or compression of ureters all cause increased EPO production by the kidneys. And lastly increased life span of RBCs has also been postulated. Most favoured of these till now is production of EPO by the

leiomyoma itself as shown by elevated levels of EPO within the uterine fibroid measured by radio-immunoassay[2] and the increased EPO level in the uterine vein as compared to the levels in the artery[3].

It has been postulated that all myomas produce EPO (with or without erythrocytosis) and the development of polycythaemia depends on whether the EPO level exceeds the normal range[1].

Diagnosis is usually made following symptoms of uterine fibroids, when a pre-op workup for hysterectomy reveals polycythaemia, or the absence of anemia despite menorrhagia in a woman having fibroids. A simple CBC will diagnose erythrocytosis. Workup includes excluding primary polycythaemia as there is a high risk of thromboembolism in this condition which should be vigilantly watched for while patients are being investigated. As seen in this case, a normal WBC, platelets and MCV value all point to a diagnosis other than primary polycythaemia. Blood volume, bone marrow examination, RBC life span and EPO levels may be used as well but are not easily available and are expensive. The clinician should keep in mind that the fibroid may be the cause of the polycythaemia and thus avoid embarking on extensive investigations. In the workup of secondary polycythaemia, the absence of a significant history, a normal chest X ray and a normal spO2 ruled out generalised hypoxia and congenital causes. The finding of normal tumour markers and absence of any other lesion on CT scan excludes para-neoplastic syndromes as a cause of the polycythaemia. Management includes repeated phlebotomies to reduce the Hb to a level appropriate for surgery, even though currently it's unclear whether secondary polycythaemia is associated with a higher risk of haematological complications during surgery (such as thromboembolism or bleeding).It has been postulated that blood should be stored for autologous transfusion during or after surgery if necessary. Post-op the Hb should return to reference range values for the diagnostic triad to be fulfilled. This may take from a few days upto 4 weeks[4]. If the post-operative Hb does not normalize then further investigations are warranted. Histopathological findings are similar to those of uterine leiomyomas.



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