East African Scholars Journal of Medical Sciences
Abbreviated Key Title: East African Scholars J Med Sci
ISSN: 2617-4421 (Print) & ISSN: 2617-7188 (Online)
Published By East African Scholars Publisher, Kenya

OPEN ACCESS

Volume-7 | Issue-1 | Jan-2024 | DOI: 10.36349/easms.2024.v07i01.002

Case Report

Myomatous Erythrocytosis Syndrome with Abnormally High Hemoglobin Level: A Case Report

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Article History
Received: 07.12.2023
Accepted: 18.01.2024
Published: 20.01.2024

Journal homepage:
https://www.easpublisher.com

Quick Response Code

Abstract: Myomatous Erythrocytosis Syndrome is a rare condition characterized by a myomatous uterus, erythrocytosis that spontaneously returns to normal red blood cells count after removal of the myomas. The etiology of this syndrome is still unknown but it has been observed that after myomectomy or hysterectomy the red blood cells count returns to normal. This case reports a 57 years old woman who presented with postmenopausal vaginal bleeding, a myomatous uterus of 16.2 x 16.1 x 22.6 cms, RBC 8.19 x 10^{12}/L, Hemoglobin 21.5 g/dL, Hematocrit 67.1% and Erythropoietin of 23 mIU/mL. She was experiencing heavy postmenopausal bleeding and lower abdominal pain such that she consented to undergo a total abdominal hysterectomy with bilateral salpingo-ophorectomy. Two weeks after hysterectomy her hematology parameters reduced to normal levels, erythropoietin level was 8.2 mIU/mL. Histology results of the uterus showed leiomyomas, all the other symptoms resolved. It is suspected that erythropoietin from the myomas was responsible for the high RBCs, and the patient did not need any other treatment.

Keywords: Myomatous Erythrocytosis syndrome, Uterine myoma, Erythropoietin.

INTRODUCTION

Uterine myomas are very common benign tumors of the female reproductive system, associated with severe menstrual bleeding and anemia. However, a few women develop myomas and have erythrocytosis. First reported by Thomson and Marson in 1953, myomatous erythrocytosis syndrome was characterized by a myomatous uterus, erythrocytosis that resolved after surgical removal of the myomas [1]. The exact pathogenesis is yet to be established, but there are several theories postulating this rare condition. Erythrocytosis can be caused by cerebellar hemangioblastoma and renal cell carcinoma, but myomatous erythrocytosis syndrome has been associated with a common and benign uterine condition, leiomyoma. Here we are presenting a case of a perimenopausal woman with huge uterine myomas and erythrocytosis.

CASE

A 57 years old woman, para 3 presented with heavy postmenopausal bleeding that was taking a longer period (7days); headache, palpitation, heaviness on the Lt side of chest, a big mass felt in the lower abdomen that was associated with periodic constipation. She had been treated in another facility and was informed she had a huge uterine myoma.

She had a long standing headache that was associated with her high hemoglobin level, and she did not get any explanations of her persistent headaches. She did not have hypertension, poor vision, dental or hearing impairments.

Physical examination showed a healthy African woman, fully conscious, not anemic and had no jaundice. The skin was clear of any rash, wounds or discoloration. Abdominal examination showed a moderate distension below the umbilicus, more on the Left side. There was a palpable solid mass, mobile with an irregular surface, it measured 12 centimeters in height in the hypogastric abdominal area. An abdominal CT scan revealed a myomatous uterus of 16.2 x 16.1 x 22.6 cms; blood work up showed RBC 8.19 x 10^{12}/L, hemoglobin 21.5 g/dL, hematocrit 67.1%, platelets and WBC were 274 x 10^{9}/L and 3.99 x 10^{9}/L respectively. *(Fig 2)* She checked

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fasting blood glucose that was 4.6 mmol/L and creatinine level was 74 µmol/L.

With the results showing a huge uterine myoma the patient was informed and given autonomy to choose between myomectomy and hysterectomy. After all considerations she decided to undergo a total abdominal hysterectomy and bilateral salpingo-ophorectomy. Preparations were done and she underwent a total abdominal hysterectomy with bilateral salpingo-ophorectomy. *(Fig. 1)* The hysterectomy tissue sample was sent for histological examination. She recovered well and a week after surgery headache, awareness of heartbeats, chest pain symptoms all subsided.

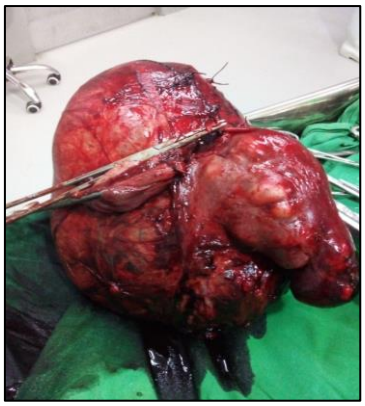
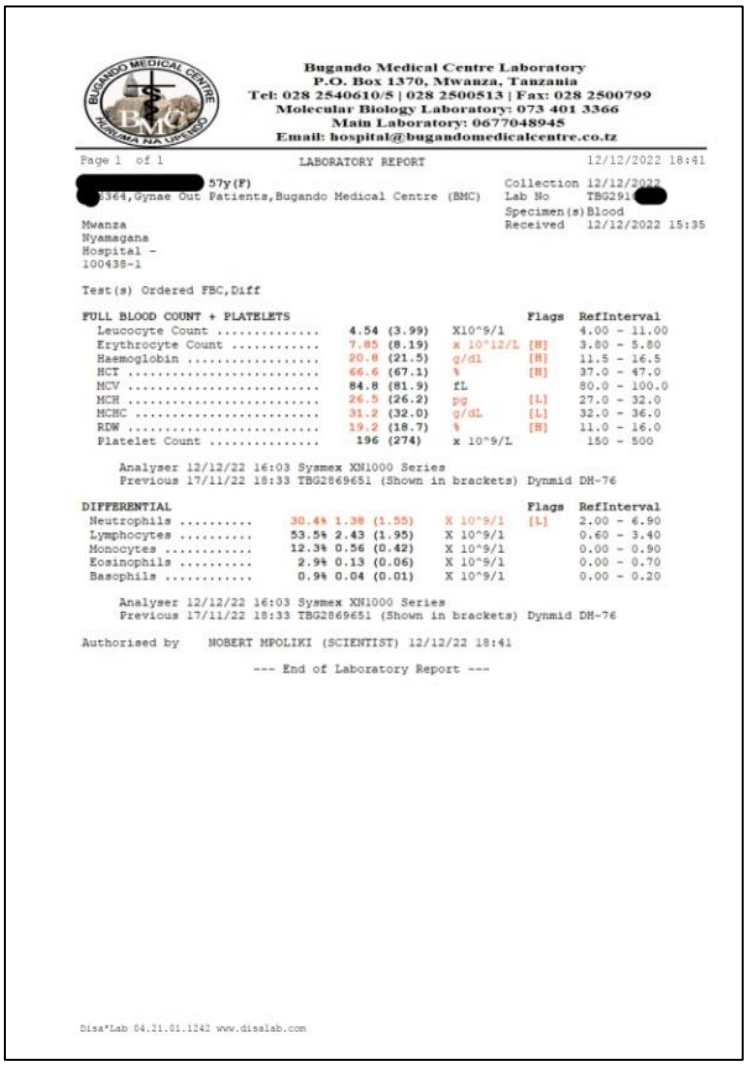


Fig 1: Uterine and ovarian samples after surgery



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Page 1 of 1 — LABORATORY REPORT — 12/12/2022 18:41

[redacted] 57y(F)
[redacted]364,Gynae Out Patients,Bugando Medical Centre (BMC)
Collection 12/12/2022
Lab No TBG291[redacted]
Specimen(s) Blood
Received 12/12/2022 15:35

Mwanza
Nyamagana
Hospital -
100438-1

Test(s) Ordered FBC,Diff

FULL BLOOD COUNT + PLATELETS			Flags	RefInterval
Leucocyte Count	4.54 (3.99)	X10^9/l		4.00 - 11.00
Erythrocyte Count	7.85 (8.19)	x 10^12/L	[H]	3.80 - 5.80
Haemoglobin	20.8 (21.5)	g/dl	[H]	11.5 - 16.5
HCT	66.6 (67.1)	%	[H]	37.0 - 47.0
MCV	84.8 (81.9)	fL		80.0 - 100.0
MCH	26.5 (26.2)	pg	[L]	27.0 - 32.0
MCHC	31.2 (32.0)	g/dL	[L]	32.0 - 36.0
RDW	19.2 (18.7)	%	[H]	11.0 - 16.0
Platelet Count	196 (274)	x 10^9/L		150 - 500

Analyser 12/12/22 16:03 Sysmex XN1000 Series
Previous 17/11/22 18:33 TBG2869651 (Shown in brackets) Dynmid DH-76

DIFFERENTIAL			Flags	RefInterval
Neutrophils	30.4% 1.38 (1.55)	X 10^9/l	[L]	2.00 - 6.90
Lymphocytes	53.5% 2.43 (1.95)	X 10^9/l		0.60 - 3.40
Monocytes	12.3% 0.56 (0.42)	X 10^9/l		0.00 - 0.90
Eosinophils	2.9% 0.13 (0.06)	X 10^9/l		0.00 - 0.70
Basophils	0.9% 0.04 (0.01)	X 10^9/l		0.00 - 0.20

Analyser 12/12/22 16:03 Sysmex XN1000 Series
Previous 17/11/22 18:33 TBG2869651 (Shown in brackets) Dynmid DH-76

Authorised by NOBERT MPOLIKI (SCIENTIST) 12/12/22 18:41

--- End of Laboratory Report ---

Disa*Lab 04.21.01.1242 www.disalab.com

Fig 2: Hematological results pre-operation

Two weeks after surgery the patient was active, felt better and the headache had resolved. Histology results revealed that she had myomas in the uterus, myoma with calcification and hyaline changes *(Fig. 3)* and ovaries had epithelial inclusion cysts and corpus albicans. *(Fig. 4)* The full hemogramme showed RBC 4.9 x 10^{12}/L, Hemoglobin 15.8 g/dL, hematocrit 39.1% and platelets 198 x 10^{9}/L and unexpectedly erythropoietin levels dropped to 8.2 mIU/mL. That is when we concluded that she had Myomatous Erythrocytosis Syndrome.

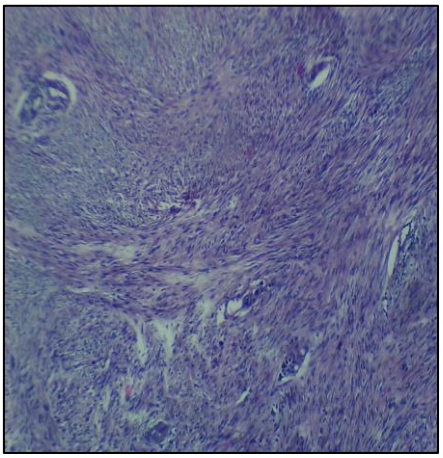
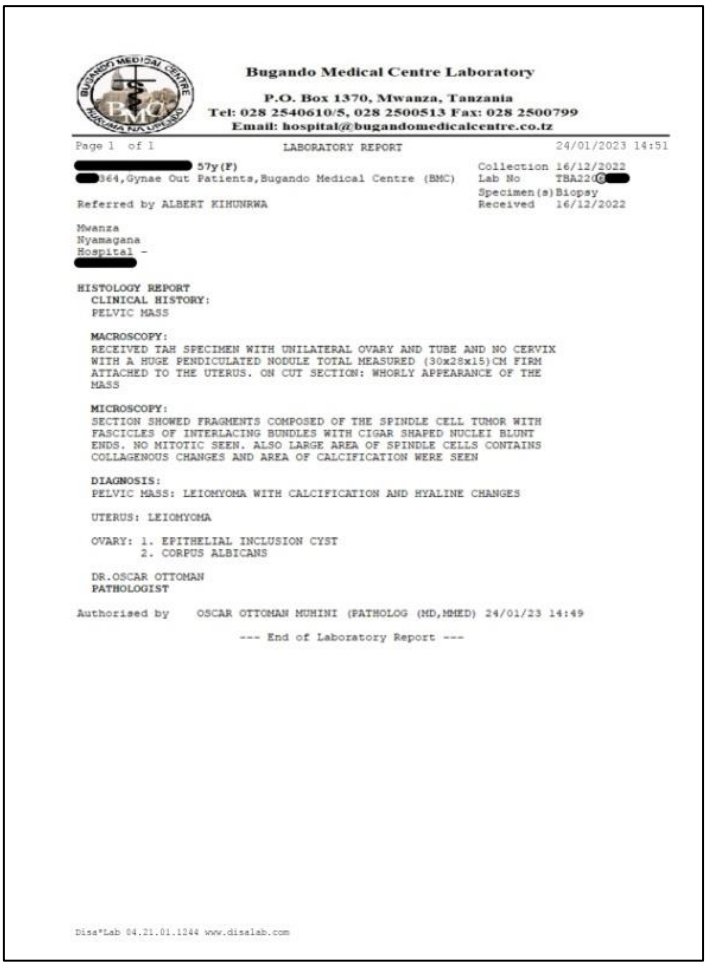


Fig 3: Histology slide magnify X100.



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[redacted] 57y (F)
[redacted]964,Gynae Out Patients,Bugando Medical Centre (BMC)
Collection 16/12/2022
Lab No TBA22[redacted]
Specimen(s) Biopsy
Received 16/12/2022

Referred by ALBERT KIHUNRWA

Mwanza
Nyamagana
Hospital -
[redacted]

HISTOLOGY REPORT
CLINICAL HISTORY:
PELVIC MASS

MACROSCOPY:
RECEIVED TAH SPECIMEN WITH UNILATERAL OVARY AND TUBE AND NO CERVIX WITH A HUGE PENDICULATED NODULE TOTAL MEASURED (30x28x15)CM FIRM ATTACHED TO THE UTERUS. ON CUT SECTION: WHORLY APPEARANCE OF THE MASS

MICROSCOPY:
SECTION SHOWED FRAGMENTS COMPOSED OF THE SPINDLE CELL TUMOR WITH FASCICLES OF INTERLACING BUNDLES WITH CIGAR SHAPED NUCLEI BLUNT ENDS. NO MITOTIC SEEN. ALSO LARGE AREA OF SPINDLE CELLS CONTAINS COLLAGENOUS CHANGES AND AREA OF CALCIFICATION WERE SEEN

DIAGNOSIS:
PELVIC MASS: LEIOMYOMA WITH CALCIFICATION AND HYALINE CHANGES

UTERUS: LEIOMYOMA

OVARY: 1. EPITHELIAL INCLUSION CYST
2. CORPUS ALBICANS

DR.OSCAR OTTOMAN
PATHOLOGIST

Authorised by OSCAR OTTOMAN MUHINI (PATHOLOG (MD,MMED) 24/01/23 14:49

--- End of Laboratory Report ---

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Fig 4: Histology results

DISCUSSION

Myomatous erythrocytosis syndrome is a rare condition that affects a few women of reproductive age. It is usually characterized by presence of uterine myoma, polycythemia or erythrocytosis and return to normal levels of erythrocyte count after removal of myomas [1].

Presence of huge uterine myoma is often accompanied by heavy bleeding and this leads to low hemoglobin (anemia); but with erythrocytosis patients present with high levels of hemoglobin [2]. In our case this was observed and our patient had a hemoglobin level of 21.5 g/dL, despite having observed prolonged episodes of pre and post menopausal bleeding.

The high levels of hemoglobin, hematocrit and red blood cells were recorded in 2 separate occasions before surgery, a typical feature of MES. Before surgery this patient was advised to donate a unit of blood to the blood bank, in anticipation that the hemoglobin will decrease to appreciable levels and relieve her of the erythrocytosis symptoms [3]. Other case reports showed a similar attempt of autologous blood collection aimed at relieving polycythemia but they did not help. She had a 1 week relief and after 1 month her hemoglobin, hematocrit and RBC levels were still high [3].

The pathophysiology of MES remains not well understood, but many hypotheses have been proposed in recent years. The most probable at present is the autonomous synthesis of erythropoetin by the myoma cells without negative feedback, concurring with Hertko who first hypothesized it in 1963 [4]. There have to be ways to evidently show presence of erythropoietin cells in the fibroids, using immuno-histochemistry histology studies. For our case we were not able to determine this, due to low resources, but strongly showed evidence of MES through autologous blood donation and resumption of erythrocytosis in 1 month, and also return to normal levels of erythrocyte count after hysterectomy [5].

Intra operative it was noted that the blood was dark red in color, and that the uterus was huge with a left broad ligament fibroid extending to the pelvic wall and compressing on the sigmoid colon, which explains why she experienced episodes of constipation. The dark red blood was caused by the abnormally high hematocrit (67.1%) and hemoglobin (21.7 g/dL). [6] Most cases showed hemoglobin of 15-17 g/dL.

In low resource setting MES remains a rare cause of secondary polycythaemia and requires the intervention of a multidisciplinary team, (gynecology, hematology and pathology teams) in order to make the diagnosis. The fact that MES is rare poses a great challenge for gynecologists in low resource setting to diagnose, and this may result in misdiagnosis and inappropriate treatment [7].

Acknowledgement

Authors are grateful to the patient who heartedly accepted publication of her case, for the benefit of science. We also appreciate the theatre and laboratory staff who expedited the preparation of the tissues for histology evaluation.

Consent to Publish

Permission to publish the case report was granted by Joint CUHAS/BMC Research and Ethical Committee. The patient consented in writing for her information to be share publically.

Author's Contribution

AK attended and operated patient. PS attended the patient after surgery. AK, RK and AKaj prepared the manuscript for the case report. SM processed the histological materials and prepared slide photos. All authors read and corrected the case report.

Conflict of Interest: All authors declare no conflict of interest. No funding was received to process this case report.

Funding

The cost for pre-operative investigations were covered by the patient, surgery costs were waived by the hospital. No funding was received from anyone to publish this case.

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Cite This Article: Albert Kihunrwa, Richard Kiritta, Prosper Shayo, Stanley Makundi, Alphaxad Kajura (2024). Myomatous Erythrocytosis Syndrome with Abnormally High Hemoglobin Level: A Case Report. *East African Scholars J Med Sci*, *7*(1), 5-8.