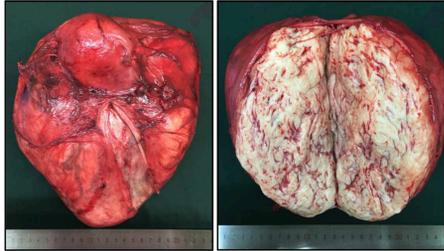
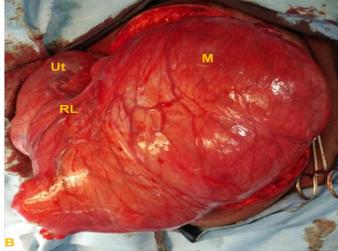


ABSTRACT

Uterine leiomyomas are one of the known tumor-associated causes of secondary erythrocytosis. Ectopic erythropoietin production by fibroid smooth muscles has been proposed and demonstrated in literature. Here, we present three cases of large leiomyomas with an incidental finding of isolated erythrocytosis on preoperative workup. Two patients underwent total abdominal hysterectomy while one patient underwent a myomectomy. Both histologic examination confirming the diagnosis of leiomyoma and serial complete blood count monitoring showing normalization of postoperative hemoglobin levels together with isolated erythrocytosis constitute the criteria to fulfill a diagnosis of myomatous erythrocytosis syndrome. All three criteria were observed in the three cases described. To date, less than 40 cases have been reported worldwide. These are the first reported cases of myomatous erythrocytosis syndrome in the Philippines.

CASES OF MYOMATOUS ERYTHROCYTOSIS SYNDROME			
	CASE 1	CASE 2	CASE 3
Age/OB Score	46/Go	45/Go	27/Go
Chief Complaint	Abdominopelvic mass	Abdominopelvic mass	Abdominopelvic mass
Other Illnesses	Hypertension Congestive heart failure Functional class I	15-year history of uninvestigated infertility	None
Abnormal Uterine Bleeding	Absent	Absent	Absent
Pelvic Examination Findings	Cervix flushed and deviated anteriorly Corpus enlarged to 26 weeks' size	Corpus enlarged to 24 weeks' size	Corpus enlarged to 24-26 weeks' size
Imaging Findings	Subserous myoma with intramural component measuring 28.2 x 22.0 x 6.3 cm, FIGO Grade 5	Subserous myoma with 50% intramural component measuring 22.7 x 21.3 x 20.2 cm, FIGO Grade 6	Intramural myoma with subserous component measuring 34.5 x 14.9 x 8.6 cm, FIGO Grade 6
Baseline Hemoglobin and Hematocrit	213 g/L 0.62	184 g/L 0.55	174 g/L 0.53
Serum Erythropoietin (Normal Range: 2.59-18.5)	29.53	24.63	Not available
Underwent Venesection	Yes	No	No
Procedure	Exploratory laparotomy, total hysterectomy with bilateral salpingectomy	Exploratory laparotomy, total hysterectomy with bilateral salpingo-oophorectomy	Exploratory laparotomy, myomectomy
Intraoperative Findings	Well-circumscribed mass at the cervicocorporeal junction measuring 18.0 x 18.5 x 12.0 cm 	Subserous, lobulated mass at the fundal area measuring 28.0 x 21.5 x 19.0 cm 	Mass at the left lateral uterine wall, subserous measuring 30.0 x 25.5 x 9.0 cm 
Biopsy Result	Leiomyoma	Leiomyoma	Leiomyoma
Post-Operative Return to Normal Hemoglobin Levels	Yes	Yes	Yes

DISCUSSION

MYOMATOUS ERYTHROCYTOSIS SYNDROME

- Isolated erythrocytosis has been shown to result from ectopic production of erythropoietin in various malignancies including renal cell and hepatocellular carcinoma. It has been found to occur in benign tumors such as cerebellar hemangioblastoma and adenomas. It is lesser known to be associated with uterine leiomyoma.
- Various mechanisms have been proposed to explain this occurrence. One hypothesis is the autonomous production of erythropoietin by the leiomyoma, which is not subjected to any negative feedback mechanism. Ectopic erythropoietin production accounts for the large sizes of leiomyoma encountered in this syndrome through its mediation of angiogenesis, mitogenesis, and inhibition of apoptosis.

PREOPERATIVE MANAGEMENT

- The etiology of isolated erythrocytosis must be identified and secondary causes identified in collaboration with hematology services. The possibility of polycythemia vera or a myeloproliferative disorder should be confirmed through Jak2 mutation testing.
- Risks for thrombosis, embolizations, and other cardiovascular complications should be considered prior to surgery. Preoperative planning should be undertaken to minimize or avoid these complications and may involve venesection as was undertaken in the first case. This may be done once or twice weekly to reduce hematocrit levels to below 0.60. Hemoglobin levels returned to normal immediately post-surgery and were maintained on subsequent outpatient visits.