



Myomatous erythrocytosis in uterine fibroid

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Abstract

Myomatous Erythrocytosis Syndrome is defined as erythrocytosis, myomatous uterus, and the return of normal hematologic values following surgical resection. The triad of a myomatous uterus, erythrocytosis, and restoration of normal hematologic parameters following surgical resection such as hysterectomy or myomectomy is characteristic of a rare entity called Myomatous Erythrocytosis Syndrome. The first reports of these conditions were by Thomson and Marson in 1953^[1].

Keywords: Non-formal education, illicit drugs, antidote

Introduction

Uterine fibroids occur in approximately 50% of women over the age of 40 years, and an estimated 50% of those are symptomatic.

Myomas can be classified as Subserosal myomas, Intramural myomas, Submucous myomas. The triad of a myomatous uterus, erythrocytosis, and restoration of normal hematologic parameters following surgical resection such as hysterectomy or myomectomy is characteristic of a rare entity called Myomatous Erythrocytosis Syndrome. The exact pathogenesis of this disease is unknown. However, there have been published case reports since then examining various hypotheses, which we will discuss here.

Case presentation

Case report

65 yr old, P3L3 with previous all normal vaginal deliveries Post Menopausal women patient came with chief complains of Pain in abdomen since 21 days.

Patient was hypertensive since one year and was taking tab. amlodipine 5 mg twice a day.

On examination

She was well built and well nourished.

BP-110/70 PR-78/MIN

CVS: S1, S2 Normal, No Murmurs

RS: AEBE,

CNS: NAD

Her per Abdomen Examination

Soft, non tender

Firm Mass palpable about 28wks size, mobile

On Per Speculum Examination

Cervix and vagina healthy

On Per Vagina Examination

Mass of 28wks size, mobile

Uterus size could not be assessed.

B/L fornices nontender

B/L adnexa not distinguished well.

Investigations

HB- 21 gm/dl

RBC- 7.3 million cells/cumm

TLC-4000 cells/cumm

PLT- 1.7 lakhs

PCV-68.9%

PBS –Normocytic normochromic

Blood Group- B positive

Urine Analysis- normal

BSL (R)-98 GM/DL

HIV, HBsAg, VDRL- negative

PT-13.5. INR-1.1

Liver function tests-WNL

Erythropoietin Level 34.30 Mu/MI

(3.7-31.5)

2D echo – normal study

USG ABD/pelvis

Uterus is grossly enlarged and shows a fibroid of size approx. 20cm x 11cm

CT scan

A large well defined, hypodense moderately enhancing solid lesion is seen arising from the fundus of the uterus, 22x12.5x17.4cm in dimensions.

MRI Pelvis

Large well defined lobulated solid mass lesion in pelvis measuring 23x 16x11cm.

Treatment

Total Abdominal Hysterectomy and Bilateral Salpingoophorectomy was done.

Intra Op findings

18x19 cm fibroid arising from fundus seen, dilated vessels were seen on the myoma.

Discussion

Thomson and Marson were the first to report erythrocytosis secondary to uterine myoma in 1953^[1] Uterine leiomyomas are benign tumours arising from smooth muscles cells of

uterus but contains varying amounts of fibrous tissues. Commonest presentation are, mass effect on pelvic organs and abdominal distortion. Problems associated with fibroids are ascites, uterine inversion, Polycythemia. Secondary polycythemia is one of the rarest association reported (0.5%). The role of erythropoietin has been suggested in the etiopathogenesis, both autonomic secretion by myomatous uterus or by the kidneys. Vascular shunts within the myoma and large uterine contribute to local tissue hypoxia which stimulates erythropoietin secretion. Menzies *et al* described compression of these uterine masses causing disruption of urinary flow leading to increased renal parenchyma pressure and erythropoietin stimulation, or even compression of renal vessels leading to renal hypoperfusion and erythropoietin secretion [2]. Paranjthy *et al* also attributed the increased size of uterine myomas to causing pressure on the diaphragm leading to hypoxia and subsequent increase in erythropoietin production [3]. This theory, however, was aborted since many patients with this condition did not show any signs of hypoxia or abnormal pulmonary function tests [4]. Production of erythropoietin happens mainly in the renal cortex and is involved in survival, growth, and differentiation of progenitor cells of the erythroid lineage. The first time the theory of uterine myomas producing excess erythropoietin was proposed in 1955 [5]. In 1968, Hertko *et al.* was the first one to discuss increased radioactive iron incorporation in a hypoxic polycythemic

mouse [6, 7]. Wrigley *et al.* later confirmed this using a 2-dose level radioactive iron assay in 1971 [8] since then, many have shown increased erythropoietin activity in uterine myoma tissue. Among them, Yoshida *et al.* demonstrated positive immune-staining for erythropoietin in the cytoplasm of leiomyoma cells [9]. Erythropoietin produced directly by uterine myoma tissue has also been demonstrated by Kohama *et al.* who used reverse-transcriptase polymerase chain reaction and Southern blot method to detect erythropoietin Mrna in myomatous tissue [5, 10]. There is some *in-vivo* experimental data to suggest that estrogen plays a role in stimulating the erythropoietin/erythropoietin-receptor signalling pathway causing increased angiogenesis, which is important in the development of this syndrome [11]. Literature review showed associated cases of erythrocytosis with leiomyoma of the esophagus and cutaneous leiomyoma, bringing up the theory of myoma cells itself being responsible for inappropriate erythropoietin secretion, regardless of its location. Apart from confirming erythropoietin production by myomatous tissue, Fabrizio Pollio and colleagues found a strong Epo-R expression in myoma endothelial cells, leading to the theory that erythropoietin production from uterine myoma not only contributes to erythrocytosis, but also stimulates proliferation and differentiation of myoblasts through autocrine and paracrine mechanisms leading to the large myomatous size almost always seen in this condition [12].

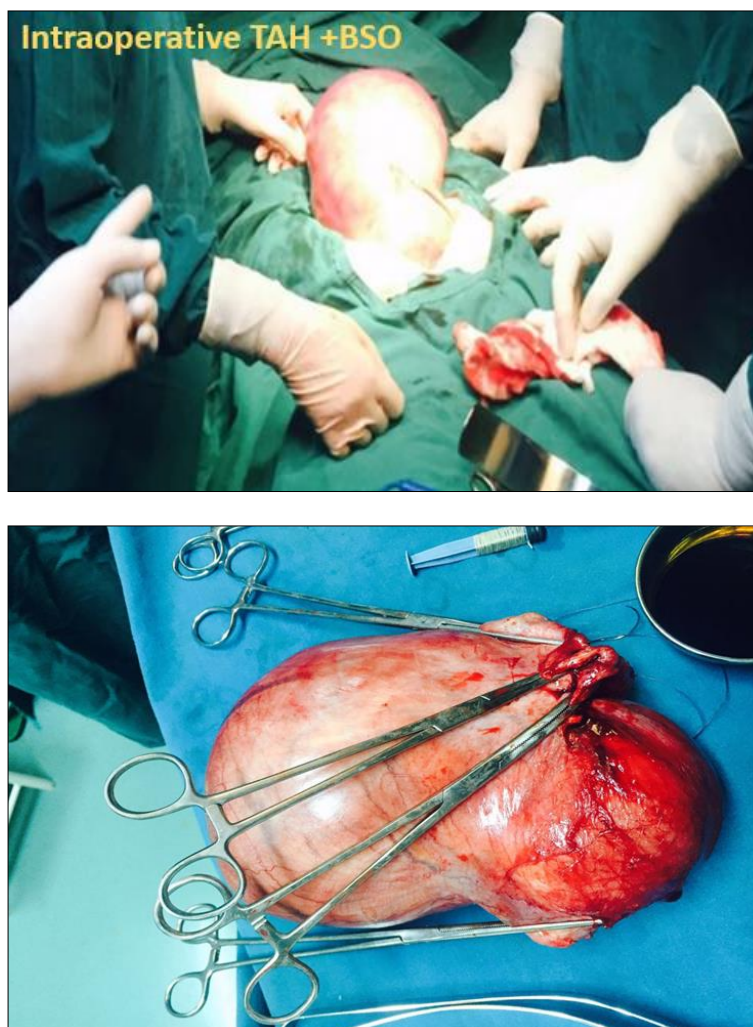


Fig 1: Specimen of uterus with cervix and fibroid

Conclusion

Uterine fibroid is one of the commonest pelvic tumours that can be associated with unusual presentation. Polycythemia is one among them. This underlines the importance of thorough physical examination including review of other systems to rule out paraneoplastic manifestation.

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