

Postmenopausal Secondary Polycythaemia due to Large Fibroid

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BACKGROUND

Uterine fibroids or leiomyomas are the most common benign uterine tumours, occurring in about 15-20% of women in their reproductive years [1]. An association of fibroids with polycythaemia has been reported in several series [2,3]. Such an association is described in the literature as Myomatous Erythrocytosis Syndrome. It is a very rare condition and, although reported first nearly seven decades ago, the aetiology is still unclear.

CASE REPORT

This case reports a 57-year old postmenopausal Caucasian lady who presented with a large pelvi-abdominal mass and a Haemoglobin (Hb) level of 23.5 g/dl, possibly the highest Hb level reported in literature secondary to fibroid uterus. She had an HCT of 69.9% and Bilirubin of 37.6 mg/dL. Both ultrasound scan and CT for abdomen and pelvis suggested a large fibroid uterus. A provisional diagnosis of Myomatous Erythrocytosis Syndrome was made, and she was considered for total abdominal hysterectomy + bilateral salpingo-oophorectomy after normalisation of her Hb and HCT. While awaiting her surgery the patient developed an episode of rectal bleeding for which she underwent both OGD and colonoscopy. She was diagnosed with Grade 2 esophagitis, large duodenal ulcer, and diverticular disease. A few weeks later, she was fit for surgery. Histology report confirmed fibroid uterus with no evidence of sarcomatous changes. Her haemoglobin remained normal 8 months later.

DISCUSSION AND CONCLUSION

An association of fibroids with polycythaemia is described in the literature as Myomatous Erythrocytosis Syndrome. Although the incidence of myomas in premenopausal women is 20 - 40%, the occurrence of the myomatous erythrocytosis syndrome ranges from 0.02 - 0.5%. It has been postulated that the real incidence of this syndrome may actually be higher as accelerated erythrocytosis may be obscured by the presence of menorrhagia [3].

In 1957, three diagnostic criteria for myomatous erythrocytosis syndrome were proposed: (1) erythrocytosis, (2) a myomatous uterus, and (3) the restoration and maintenance of normal haematological values after hysterectomy [4]. Our case not only fulfilled these criteria but also reported the highest level of Hb and HCT reported in literature. This case report also presents large duodenal ulcers as one of the most serious complications of polycythaemia.

Despite the fact that the syndrome was first reported in 1953 by Thomson and Marson [5], the aetiology is still unclear. There are, however, several mechanisms that have been proposed. Ectopic production of erythropoietin by the leiomyoma is the most favoured aetiopathogenesis [6]. Ectopic erythropoietin production is known to occur as a complication of various tumours such as renal cell carcinoma, hepatocellular carcinoma, and cerebellar hemangioblastoma. However, it is less well known that it can also occur as a complication of uterine leiomyoma [7].

Several studies using immunostaining techniques have confirmed the production of erythropoietin by the myoma tissue [6]. Many studies, through different approaches, have shown increased erythropoietin activity in uterine myoma tissue. Literature review showed associated cases of erythrocytosis with leiomyoma of the oesophagus and cutaneous leiomyoma, bringing up the theory of myoma cells themselves being responsible for inappropriate erythropoietin secretion, regardless of their location [8].

Gonadotrophin-releasing hormone (GnRH) analogues are commonly used preoperatively to control anaemia secondary to menorrhagia or shrink uterine fibroids to facilitate surgery. There is no published association between the use of GnRH analogues and hyper erythropoietinaemia [6]. As in our case report, removal of the tumour was followed by remission in almost all cases, so far reported. Alternative treatment options such as phlebotomy will normalise Hb and HCT for a short time.

CONCLUSION

Myomatous Erythrocytosis Syndrome is a rare presentation of uterine fibroids, as most women present with menorrhagia and/or anaemia. It is important however, to recognize the rare association between polycythaemia and fibroids to minimize unnecessary investigations. In the clinical setting of polycythaemia, it is imperative to rule out polycythaemia vera that does not only lead to serious, life-threatening thrombosis but also has different treatment than myomatous erythrocytosis syndrome, which resolves after surgical removal. Costly investigations however, especially genetic testing, should be reserved for persistent polycythaemia following hysterectomy/ myomectomy. The exact role of erythropoietin in disease pathogenesis is unknown. As a single factor cannot be accepted in all cases, the aetiology seems most likely multifactorial. Moreover, clinical expression of the syndrome is also poorly understood; while we report the highest Hb level in literature of 23.5 g/dl with only a 3 kg fibroid, Abdul Ghaffar, et al (2008) reported a bigger fibroid of 5.05 kg with an Hb of only 22.1 g/dl [9]. The high frequency of uterine fibroids in women throughout their life does not explain the rarity of this Myomatous Erythrocytosis Syndrome. Together with the uncertainty of the aetiology, more research is needed in the future to shed a light on this rare condition and its aetiology.

REFERENCES

