

Case Report

Myomatous erythrocytosis syndrome: a case report in Ogbomoso, Southwest Nigeria

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متلازمة كريات الدم الحمراء ميوماتوس: تقرير حالة في أوغبوموسو، جنوب غرب نيجيريا

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ملخص

الأورام الليفية الرحمية هي الأورام الحميدة النسائية الأكثر شيوعاً. خلايا العضلات الملساء في عضل الرحم هي المسؤولة عن تشكيل الأورام الليفية الرحمية. تحدث في أكثر من 50٪ من النساء فوق سن 35 عاماً، مع أكثر استعداداً في نيجرويدز من القوقازيين. متلازمة كريات الدم الحمراء ميوماتوس التي تتميز بوجود الورم الليفي كبير مع كثرة الحمر وبعض من المظاهر السريرية للبوليسثيميا مثل آلام الرأس، وضوح الرؤية والحكة والجلطات الدموية الذي يحل تماماً بعد استئصال الرحم. الحالة المعروضة هنا هي فقرة 4 + 47 عاماً التي كان لها الأورام الليفية الرحمية ضخمة وكشف تقييم آخر أيضاً أن المريض كان كثرة الحمر. تم تقييمها من قبل أخصائي أمراض الدم، وكان في نهاية المطاف استئصال الرحم الكلي في البطن و ساليبنغوفوركتومي الثنائية بسبب تاريخ عائلي إيجابي من سرطان المبيض. لها الشخصية الدموية وشكوى مشتركة من الصداع حلها بشكل كبير بعد الجراحة.

Abstract

Uterine fibroid are the most common gynecological benign tumors. The smooth muscle cells of the myometrium are responsible for the formation of uterine fibroid. They occur in more than 50% of women over the age of 35 years, with more predispositions in the Negroids than the Caucasians. Myomatous erythrocytosis syndrome which is characterized by presence of large fibroid with polycythaemia and some of the clinical features of polycythaemia like head ache, blurring of vision, pruritus and thromboembolism which resolves completely after hysterectomy. The case presented here is

a 47-year-old para 4+1 who had huge uterine fibroid and further evaluation also revealed that patient had polycythaemia. She was evaluated by the haematologist and eventually had a total abdominal hysterectomy and a bilateral salpingoophorectomy because of the positive family history of ovarian cancer. Her haematologic profile and her common complaint of headache resolved significantly following the surgery.

Keywords: Myomatous, erythrocytosis, Nigeria.

Introduction

Uterine fibroid are the most common gynaecological benign tumours arise from the smooth muscle cells of the myometrium.⁽¹⁻³⁾ The pathogenesis of fibroids is still unclear.⁽³⁾ Uterine fibroids are found in more than half of women over age 35 years.⁽¹⁻⁴⁾ Myomatous

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erythrocytosis syndrome is said to occur when there is large fibroid causing increased erythropoietin resulting in polycythaemia which may be accompanied by suggestive features like headache, blurring of vision, pruritus and thromboembolism which are as a result of the polycythaemia and resolves completely after hysterectomy.

Case report

A 47-year-old para 4+1 whose last confinement was 10 years before presentation in the clinic with a history of abdominal swelling of three years duration. The swelling has been gradually increasing in size and menorrhagia but no associated pain. There were no associated urinary or gastrointestinal symptoms. There were occasional episodes of headache, no associated history of trauma; there was a family history of ovarian cancer. On clinical examination, she was not pale, not jaundiced and there was no bilateral pedal swelling. The Abdomen was enlarged and moved with respiration, the abdominopelvic mass was similar to that of a 36 weeks gestation (Fig. 1).



Figure 1: Patient in supine position before the commencement of surgery.

The liver, spleen and kidneys were not palpably enlarged. Others systems were essentially normal. Investigation results include packed cell volume 56%, bone marrow aspirate result excluded the likelihood of primary cause of the polycythaemia. Ultrasound report was in keeping with a huge uterine mass probably a fibroid with evidence

of dilatation of the calyces worse on the right. Computer tomography scan could not be done due to financial constraint, other investigation reports were essentially normal. She was counselled for a total abdominal hysterectomy and bilateral salpingoophorectomy for which she consented. A review was done by the haematologist prior to the surgery. Thromboprophylaxis was ensured prior to surgery and patient had venesection done to further reduce the risk of thromboembolism. At surgery, a huge uterine mass with fibroid masses were removed (Fig. 2). Her post-operative recovery was satisfactory and her postoperative packed cell volume was 30%. The usual complaint of headaches had resolved significantly. Her haematologic profile was followed up for up to one year post surgery and was found to have remained in the normal range.

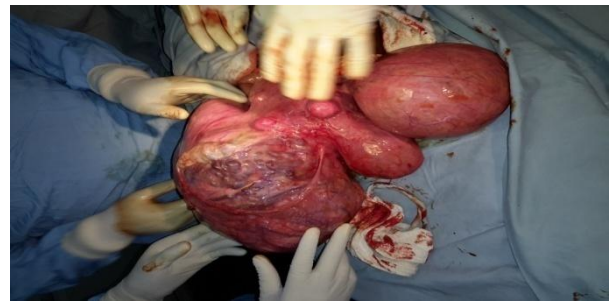


Figure 2: Uterus showing the multiple fibroid masses.

Discussion

Uterine fibroids are the most common benign tumors of uterus; it is also a common finding in cases of female pelvic tumours. They are usually asymptomatic and when these symptoms occur they vary from abnormal uterine bleeding, mass effect on the pelvic and abdominal organs, and occasionally abdominal swelling. Literature search shows that rarely polycythaemia may be associated with uterine fibroid⁽⁵⁻⁶⁾. This condition increases the viscosity of the blood and causes symptoms like head ache, pruritus, visual disturbance and venous thrombosis. When this occurs it is pertinent to involve the haematologist to be able to exclude other medical conditions like myeloproliferative disorders⁽⁷⁾.

Three theories probably explained the condition known as myomatous erythrocytosis syndrome which is characterized by presence of large fibroid with polycythaemia and some of the clinical features of polycythaemia like headache which resolves completely after hysterectomy⁽⁷⁾. These theories include presence of hypoxia suggesting shunting within the tumour, second, the uterine fibroid was compressing the ureters resulting in inappropriate excessive production of erythropoietin by the kidneys, and third, the tumour itself may have been producing the erythropoietin. In the case of the case presented, there was calyceal dilatation which suggests the second postulate. However, there was a limitation checking for the third postulate due to the unavailability of the facility to radioimmunoassay for erythropoietin⁽⁸⁾. A recent case report was published providing

further evidence for in vivo erythropoietin production by the myomatous uterus where the erythropoietin levels in the uterine vein and artery were measured showing large gradient⁽⁸⁾.

There should always be a high index of suspicion when a patient presents with a uterine fibroid because the diagnosis of myomatous erythrocytosis syndrome may be missed especially if the patient has associated menorrhagia.

In conclusion, uterine fibroid is a common pelvic tumour that can have rare associations like polycythaemia and its attendant problems. Therefore, the approach to the management of myomatous erythrocytosis syndrome must be multidisciplinary.

Conflict of Interest: None.

Ethical Approval: Patient's consent obtained.

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