Paraneoplastic Erythrocytosis in a Malignant Ovarian Steroid Cell Tumour

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Abstract Neoplastic polycythemia is a condition in which high hematocrit levels result from a neoplastic lesion in the body. It is a rare condition and occur because of excess erythropoietin or androgen production by the neoplastic cells. Steroid cell tumours of the ovary though notorious for producing endocrine changes in the body have rarely been associated with polycythemia. We present the case of a post menopausal lady who developed polycythemia and features of virilisation. Further investigations revealed that she had steroid cell tumour of the left ovary. Her signs and symptoms resolved after surgical removal of the tumour.

Keywords Polycythemia · Ovarian tumour · Malignancy · Erythrocytosis

Introduction

Steroid cell tumours of ovary are rare and constitute 0.1 % of all ovarian tumours [1]. They generate interest because they are often associated with paraneoplastic and endocrine changes in the patients [2]. Paraneoplastic erythrocytosis is a term used when tumours produce either erythropoietin or

androgen which in turn stimulates excess erythropoiesis [3]. We report a case of a post-menopausal woman who presented with erythrocytosis due to steroid cell tumour of ovary not otherwise specified (NOS). Her symptoms and signs improved after surgical removal of the tumour.

Case Report

A 55-years-old lady (para 3, live 3) was seen with complaints of excessive hair growth on face, chest & limbs, hair loss from scalp and deepening of voice of 2 months duration. She was also hypertensive and diabetic with her hemoglobin values in the range of 19–20 gm/dl before presenting at our hospital. She also had non-significant appetite and weight loss. On further questioning in our clinic, she denied history of any drug or hormonal intake. She had attained menopause at the age of 47-years. Before that her menstrual cycles were normal and regular. Examination showed that her conjunctive was suffused and face was plethoric. She had male type baldness, excessive hair growth on the face, chest and abdomen. Her blood pressure was 130/80 on medication. External genital examination showed clitoromegaly. Vaginal examination showed a normal cervix and vagina. Bimanual examination revealed bulky uterus and an adnexal mass on the left side. Investigations were as follows: Hemoglobin 19.5 gm/dl, Hematocrit 55 %, WBC and platelets normal. Fasting plasma glucose was 146 mg/dl and 2 h post lunch was 176 mg/dl. Liver and renal serum chemistries were normal. Hormonal levels were as follows: Serum testosterone >1,500 ng/ml (normal 6–82), dehydroepiandrosterone 205 µg/dl (18.9–205). Serum prolactin and thyroid hormones were within normal range. Serum CA levels were elevated at 1,413 IU/ml (0–35), whereas carcinoembryonic antigen and alpha

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fetoproteins were normal. PAP smear was atrophic. Findings on transvaginal ultrasound examination were: endometrial thickness of 7 mm, left adnexal mass 5×5 cm with ill defined margins and presence of ascites. A contrast enhanced CT scan of the abdomen confirmed the findings seen on ultrasound examination with normal adrenal glands and no additional tumour or metastasis anywhere else in the body. Ascitic fluid analysis for malignant cytology was negative. Endometrial aspiration revealed benign glandular epithelial cells along with few macrophages. With a provisional diagnosis of androgen producing ovarian tumour, patient underwent exploratory laparotomy. Left ovary was replaced by 8×10 cm necrotic tumour and there was a breach in the capsule. Right ovary and peritoneal surface were normal. Uterus was of 6-weeks size. One litre of ascitic fluid was taken out, malignant cytology of fluid was negative again. Total abdominal hysterectomy with bilateral salpingo-oophorectomy and infracolic omentectomy was carried out. Multiple peritoneal biopsies were done. Histopathological examination of the tumour findings were consistent with ovarian steroid cell tumour NOS. Peritoneal biopsies did not show tumour infiltration. FIGO staging was IC. Post operative patient recovered uneventfully. Her hemoglobin levels varied between 12 and 14 gm/dl in the first postoperative week before stabilising to 12 gm/dl at discharge from the hospital. The hirsutism and other virilisation features also improved gradually over a period of 6-weeks. The tumour was considered malignant and patient received four cycles of chemotherapy with paclitaxel and carboplatin. On a follow-up period of 18 months, there was no evidence of recurrence of the tumour by ultrasound and her hemoglobin remained stable at 12.5 gm/dl.

Discussion

Previously known as “Lipid cell tumour” or “Lipoid cell tumour”, the term steroid cell tumour was coined by Hayes and Scully to reflect accurately the morphological appearance of these tumours [1]. Ovarian tumours are grouped as: stromal luteomas, Leydig cell tumours (hilus and non-hilar type) and steroid cell tumours NOS. Steroid cell tumours that cannot be diagnosed specifically as stromal luteomas or Leydig cell tumour subtypes are classified as steroid cell tumours, NOS and constitutes 60 % of steroid cell tumours [2]. Their presentation in patients is generally related to signs and symptoms of androgen, oestrogen or rarely progesterone excess [1].

In our patient, excess production of testosterone can explain all the clinical features. Receding hairline, male-type baldness, facial and body hirsutism were related to hyperandrogenism. The biochemical abnormalities in the form of polycythemia, insulin resistance, mild hypertension

were also related to excess testosterone production. The improvement in virilizing features, return of hematocrit to normal value and normalisation of blood pressure and sugar after removal of tumour confirms the source of excess testosterone towards the ovarian tumour [4, 5].

Our patient received post operative chemotherapy as it was felt that the tumour was malignant. The features supporting malignant nature of the tumour were size of the tumour more than 7 cm, breach of the capsule, presence of necrosis in the tumour and ascites [3]. In contrast to benign nature of both the stromal luteomas and Leydig cell tumours, 25–43 % of steroid cell tumours, NOS are generally malignant [2]. The treatment consists of total abdominal hysterectomy with bilateral salpingo-oophorectomy in a woman who has completed her family. Conservative surgery with unilateral oophorectomy can be done in a woman who desired future pregnancy. The chances of tumour occurring bilaterally are approximately 6 % and thus a close follow-up can be done while conserving one ovary. The best predictors of malignant nature of the tumour are presence of two or more mitotic figures per ten high power fields, grade 2–3 nuclear atypia, necrosis and tumour size of more than 7 cm [2].

Our patient had some unique features in the form of presentation with polycythemia and presence of ascites [3, 4]. Both features have been described very rarely in otherwise rare malignant ovarian steroid cell tumour not otherwise specified. A high-index of suspicion in a female patient presenting with above mentioned features would help in early diagnosis. Erythropoietin assay if available should be done in all cases as this will support the view that the ectopic production of hormone from the tumour. This was not done in the present study and this is one of the limitation of the present study. The prognosis of steroid cell tumours of the ovary is good even if the tumour is malignant.

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