Case Report

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A Rare Case of Malignant Ovarian Tumor with Para neoplastic Syndrome

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Abstract

A 17 year old girl was admitted with lump in abdomen, dyspnoea, secondary amenorrhoea and loss of appetite since 4 months and swelling over right arm, shoulder and right aspect of neck since 15 days. Ultrasound and CT abdomen and pelvis showed malignant mass lesion of adnexa with ascites. Doppler of Right arm showed thrombus in Right subclavian vein. Her coagulation profile was altered with INR of 1.4. Considering possibility of germ cell tumor LDH, β HCG and Alpha fetoprotein were done. All these markers were raised. She was started on low molecular weight heparin in view of subclavian vein thrombosis. Neoadjuvant chemotherapy was given followed by staging laparotomy. Histopathology report was suggestive of malignant mixed germ cell tumor. Postoperatively she received 3 cycles of chemotherapy. Main aim of publishing this case is presence of Para neoplastic Syndrome.

Keywords: Adolescent Ovarian Tumor; Germ Cell Tumor; Para Neoplastic Syndrome; Subclavian Vein Thrombosis; Neoadjuvant Chemotherapy.

Introduction

Germ cell tumors are second amongst all ovarian cell tumors. Germ cell tumors arise from primitive germ cells of ovary. They account of 20-25% of all benign and malignant tumors of ovary [1]. They are common in young girls and women of reproductive life. There is role of neoadjuvant chemotherapy in management of malignant germ cell tumors which are in advanced stage or inoperable. Paraneoplastic syndromes are systemic manifestations of cancer that are not due to direct (local or metastatic) effects of the tumor. Various paraneoplastic syndromes may occur in association with ovarian tumors, especially epithelial ovarian cancers. These syndromes include nervous system disorders (cerebellar degeneration, polyneuritis), connective tissue disorders (dermatomyositis), hematologic disorders (hemolytic anemia, disseminated intravascular coagulation), cutaneous disorders (acanthosis), and nephrotic syndrome [2]. Here with reporting a case of malignant mixed germ cell tumor with Para Neoplastic Syndrome in a 17 year old girl. She was managed with neoadjuvant chemotherapy with low molecular heparin because of poor general condition and then staging laparotomy was done.

Case

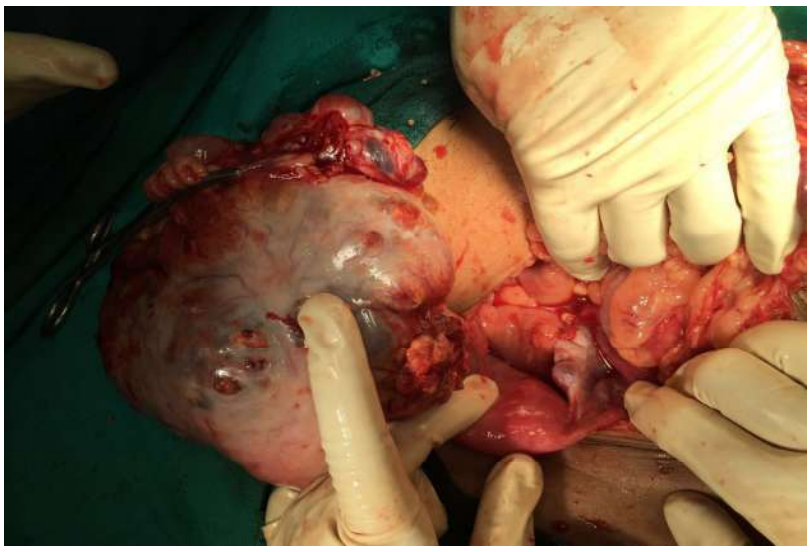
Miss AM a17 year young girl was admitted with complaints of distension of abdomen, lump in abdomen, dyspnoea, secondary amenorrhoea and loss of appetite since 4 months. She also had history of swelling over right arm, shoulder and right aspect of neck since 15 days She was admitted for

15 days at a private hospital and was referred after 15 days of hospitalization. She attained menarche at the age of 14 years with eumenorrhic cycles. It was first episode of secondary amenorrhoea. There was no family history of malignancy.

Her general condition was poor. She had tachycardia with pulse rate of 120/min and tachypnea with respiratory rate of 32/min. She was grossly pale. There was no icterus, cyanosis or lymphadenopathy. She had swelling over right arm and shoulder. Her breast examination, cardiovascular, and respiratory examination revealed no abnormality. Abdomen was tense and a mass of 32 wks pregnant uterine size was palpable which was having variegated consistency with irregular surface. It was immobile and non-tender. Per rectal examination confirmed abdominal findings and uterus was normal size. Provisional diagnosis of ovarian tumor was made and she was investigated. Her haemoglobin was 5.2gm%. Peripheral smear was suggestive of dimorphic anaemia. Thyroid function test, Kidney function test, Liver function test, Antiphospolipid antibody and homocysteine were normal. Ultrasound pelvis was suggestive of neoplastic mass lesion in pelvis ?ovarian mass with mild to moderate ascitis. CT abdomen and pelvis showed malignant mass lesion of adnexa with ascites. Uterus was normal. Doppler of Right arm showed thrombus in Right subclavianvein.Her coagulation profile was altered with INR of 1.4. Serum LDH, β HCG and Alpha fetoprotein were increased with LDH level 2466 units/ml (Normal<248 units/ml). Alpha fetoprotein level 2015ng/ml (Normal- 8-89 Ng/ml). β HCG level17596 mIU/ml. Ca 125 was normal (5mIU/ml)

She was diagnosed as a case of malignant mixed germ cell tumor with Para Neoplastic Syndrome with Subclavianvein (deep vein) thrombosis with severe anaemia and cardiorespiratory embrassement. Three units of packed red cells were given. Enoxaparin (Low molecular weight heparin) 40 mg twice a day was given for seven days in view of right subclavian vein thrombus and raised INR. Therapeutic paracentesis was done. Without waiting for cytology report, Neoadjuvant chemo therapy was initiated on basis of raised tumor markers as she was unfit for staging laparotomy. She was started with BEP (Bleomycin, Etoposide and Cisplatin) regime and first cycle dose was given 25% of the calculateddose in view of poor general condition. She tolerated this dose of chemotherapy and was given 3 cycles of Bleomycin, Etoposide and Cisplatin. Cytology was suggestive of germ cell tumor. Her menstrual cycles resumed after first cycle of chemotherapy. There was dramatic improvement in general condition with significant improvement in appetite. Swelling in Right arm, shoulder and right aspect of neck disappeared with normalization of INR. (0.9) Doppler showed disappearance of thrombus in subclavian vein and there was drastic reduction in ascites. At the end of three cycles of chemotherapy size of ovarian mass reduced from 32 weeks to 20 weeks. Tumor markers were decreased dramatically with serum level of LDH 900 units/ml, Alpha fetoprotein 1020ng/ml and βHCG 9000mIU/ml.

Staging laprotomy was done. Intra-operatively (as shown in photograph 1) there was minimal ascites with presence of right sided mass of 15x12x10 cm



Photograph 1: Laparotomy findings showing Right sided ovarian tumor with ruptured capsule and left sided normal size ovary.

with varigated appearance and capsule ruptured. Tumor deposits were present on posterior surface of uterus, Pouch of Douglas and uterovesical fold of peritoneum. Other ovary was normal but omentum had tumor deposits. Paraaortic lymph nodes were not enlarged. Fertility conserving surgery was done with Right sided Salpingo-ovariotomyalong with infracolic omentectomy. Biopsy was taken from peritoneal deposits. Histopathology was suggestive of malignantmixed germ cell tumor. Peritoneal fluidcytology, peritoneal biopsy and omentum showed presence of malignancy Hence, surgical staging was stage III. Her postoperative period was uneventful. She received 3 cycles of BEP postopertively. Tumor markers were repeated andwere normal. Presently, she is on regular follow up.

Discussion

In first two decades of life almost 70% of ovarian tumors are germ cell origin and 1/3rd of these are malignant [1]. Malignant mixed germ cell tumors compromise only 2-5% of all ovarian tumors. In contrast to epithelial tumors they grow very fast. Most common mixed malignant germ cell tumorsare dysgerminoma (80%), endodermal sinus tumor (70%) and immature teratoma (53%) as reported in a series [3]. Embryonal carcinoma, choriocarcinoma and polyembryoma are rare type of germ cell tumors. Malignant mixed germ cell tumor is a type of tumor that consists of two or more malignant germ cell components. Tumor markers such as alpha fetoprotein (AFP), Beta human gonadotrophin (βHCG) and Lactic dehydrogenase (LDH) will help in diagnosis, prognosis and follow up of these tumors. As these tumors are common in younger age group fertility conserving surgery is always the mode of management. The best part is they are chemosensitive. Many cases of malignant mixed germ cell tumors are reported inthe literature [4,5,6,7,8]. Literature also mentions cases of malignant germ cell tumors which are managed by neoadjuvant chemotherapy [9], but in literature there is no mention of paraneoplastic syndrome with germ cell tumor. There is one case report in the literature of Meig's Syndrome which was associated with axillary vein thrombosis [10].

Aim of publishing this case isthere is no mention of paraneoplastic syndrome with germ cell tumor in literature.

To Summarize,

1. Multidisciplinary approach is required in cases of complicated malignant ovarian tumor in adolescent age group.
2. Neoadjuvant chemotherapy can be initiated on basis of raised tumor markers in malignant germ cell tumors.
3. Neoadjuvant chemotherapy leads to better cyto-reduction and reduction in intraoperative and post-operative morbidity and mortality

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Disclosure

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