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Case Report

Ewing sarcoma concomitant with pregnancy: A case report

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Abstract

Cancer coexisting with pregnancy is rare and presents considerable implications for the patient and the fetus. Malignant bone tumors are rare with Extra-skeletal Ewing sarcoma being a rare subtype amongst the Ewing sarcoma family of tumors (ESFT). Development of such a tumor in pregnancy is still rarer. Management strategies in such rare instances are based on anecdotal evidence and a limited number of available case reports and series. Here we report a case of a pregnant patient presenting with extra-skeletal Ewing sarcoma in third trimester of pregnancy.

Keywords: Pregnancy, Ewing sarcoma, Tumor.

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1. Introduction

Coexistant cancer with pregnancy is rare with an incidence of 1 case per 1000 deliveries.[1,2] Malignancy has considerable implications for the patient and the fetus. Melanoma is the most common cancer reported with pregnancy followed by cervical and breast cancer. Malignant bone tumors are rare, accounting for only 0.2% of all malignancies, commonest being osteosarcoma, Ewing's sarcoma chondrosarcoma. Extra-skeletal Ewing sarcoma is a rare subtype amongst the Ewing sarcoma family of tumors (ESFT). It is a highly aggressive soft tissue tumor that responds considerably well to local therapies like complete surgical resection. The development of extraskeletal subtype is rare and such a development in the context of a pregnancy is still rarer. In view of its scarce nature, there is scant evidence available to the clinicians to guide in the management of the disease. Therefore, management strategies are based on anecdotal evidence and a limited number of available case reports and series. Here we report a case of a pregnant patient presenting with extra-skeletal Ewing sarcoma in third trimester of pregnancy.

2. Case Report

A 24-year-old lady, G2A1 (second gravida with previous one abortion) presented to the labor room at 35 weeks gestation with lump in the inguinal region in labor. Patient had a complaint of progressive pain in left inguinal region since 4 months. The pain radiated to the left lower leg and was associated with swelling in the inguinal region and difficulty in walking. On examination, a lump around 30×15 cm in size was appreciated in the left inguinal region and was red, indurated, tender and highly vascular. Patient had sought a consultation with private practitioner prior to presenting to our hospital where an ultrasonography of the left lower limb was done and was suggestive of a complex mass of the iliopsoas region with enlarged left inguinal lymph node. A Fine Needle Aspiration Cytology (FNAC) was performed which depicted round cell sarcoma likely Ewing's. As the patient had presented in active labor, and no imaging pertaining to the extent of growth was available, there was a dilemma regarding the mode of delivery. The patient was already 5 cm dilated at admission and the pelvis seemed adequate. So, a decision for vaginal delivery was taken with a plan for classical cesarean section if need for operative delivery arose. The labor progressed smoothly, and the

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patient delivered an alive baby girl weighing 1.78 kg with good Apgars. The immediate postpartum period was uneventful. X-Ray of left thigh with hip and MRI Pelvis was done in consultation with the orthopedics department of the hospital. MRI pelvis showed an enhancing mass of 35 cm in the pelvis, extending from the left presacral region to the left thigh with no bone involvement. The previous FNAC slides were reviewed and the diagnosis of Ewing's Sarcoma was made. After a thorough search for metastasis and patient counselling, chemotherapy was planned by the radiotherapy department. Patient received 6 cycles of chemotherapy consisting of alternate cycles of Vincristine, Adriamycin, Cyclophosphamide on day 1 followed by Ifosfamide and Etoposide combination for 3 days every 21 days. Post neoadjuvant chemotherapy, the tumor did not seem surgically resectable so radiotherapy, 28 cycles of 50.4 Gy were given. There was gross reduction in the size of tumor and the patient is on follow up.

3. Discussion

A pregnancy presenting with a concomitant malignancy presents unique challenges to the treating obstetrician in terms of decision to treat the malignancy, the mode of treatment, and gestation for termination of pregnancy and mode of delivery. Our patient presented with extra-skeletal Ewing sarcoma, which, amongst the four sub types within the high grade and aggressive EFST, is rare.[3] Unlike the skeletal sarcomas presenting in children and adolescent males, the extra skeletal subtypes occur in soft tissue with lesser male preponderance and increased incidence in second and third decade of life. The subtypes arise from a common gene translocation in a fused EWS/FLI-1 gene, so the treatment of all subtypes is thus similarly based.[3,4] Standard treatment consists of neoadjuvant chemotherapy followed by definitive resection surgery or radiation therapy to the primary site. The usual chemotherapeutic agents used are vincristine, doxorubicin, ifosfamide, etoposide and cyclophosphamide in various combinations. Neoadjuvant chemotherapy is given keeping micrometastasis in mind. Molecular targeted therapy for treatment is under investigation. ESFT are known to respond dramatically to initial therapy but have a propensity to recur. PET/CT helps in post therapy surveillance and is useful in detecting both local and distant metastasis.[5-8]

Table 1 represents the literature review of various patients presenting with Ewing sarcoma, both skeletal and extraskeletal variants. Most of the cases were treated during the pregnancy with chemotherapy with no obvious effect on the fetus. Of the 19 cases reported in literature, 10 were treated with chemotherapy during the course of pregnancy, while 2 each received radiotherapy and surgical management. Only one patient out of the 10 treated with chemotherapy during pregnancy had an intrauterine demise of the fetus at 23 weeks.[24] Medical termination of pregnancy was done for two patients to initiate chemotherapy. Dactinomycin, cyclophosphamide, bleomycin, vincristine, doxorubicin and ifosfamide are the various chemotherapeutic agents which have been used for the disease during the pregnancy. Our patient presented to us at 35 weeks gestation in labor, so the chemotherapy was commenced in the postpartum period.

With no imaging available, the extent of the tumor was not defined, thus there was a difficulty in deciding the mode of delivery. There was a doubt that the tumor may have extended to the pelvic cavity and may impede the progression of labor. However since the pelvic examination seemed favorable, so a trial of vaginal delivery was given and the patient delivered vaginally. As per literature, most of the patients with pelvic or proximal femoral masses have been delivered by cesarean section. Vaginal delivery in our patient may be explained by extraskeletal nature of the disease confined to the left inguinal region with no pelvic extension.

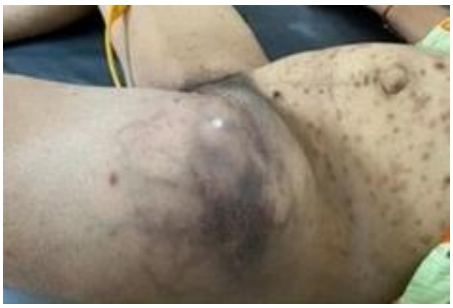

Figure 1: Patient with lump on left inguinal region on admission

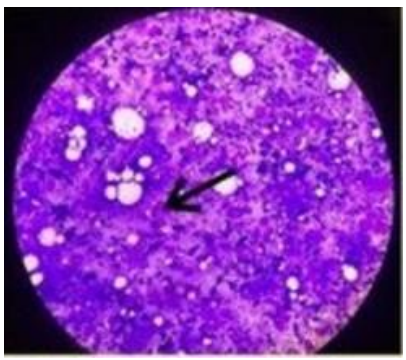

Figure 2: Single scattered population of round-oval cells, with scant cytoplasm

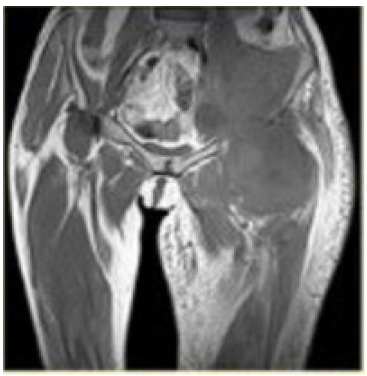

Figure 3: MRI (Stir) Coronal section after five cycles

Table 1: Literature review of patients who have presented with Ewing sarcoma with pregnancy

S. No.	Author	Gestational age at presentation	Presenting symptoms	Site of primary tumor	Evidence of metastasis	Treatment prior to delivery	Treatment after delivery	Mode of delivery	Gestation at delivery	Outcome (maternal)	Outcome (fetal)
1.	Lysyj et al 1963[9]	32 weeks	Pain in right leg	Right superior pubic ramus (skeletal)	Right lung base and right parietal bone of skull	-	Radiotherapy, Chemotherapy (5-Flourouracil)	Cesarean section	36 weeks	Died 4 months after delivery	Normal
2.	Greenberg et al 1982[10]			Left tibia (skeletal)	Mid-occipital mass, Left lung nodule, Placental nodule	Radiotherapy	Radiotherapy, Chemotherapy	Vaginal delivery		Died 7 weeks after delivery	Normal
3.	Gililland & Weinstein 1983[11]			Femur (skeletal)	None	Radiotherapy, Chemotherapy	Radiotherapy, Chemotherapy	Cesarean section		No evidence of disease 30 months after delivery	Normal
4.	Simon et al 1984[12]			Distal femur (skeletal)	None	-	Radiotherapy, Chemotherapy after abortion	Abortion		No evidence of disease 3 months after delivery	-
5.	Haerr & Pratt 1985[13]	25 weeks		Left iliac bone (skeletal)	None	Chemotherapy (dactinomycin, cyclophosphamide, bleomycin, vincristine, doxorubicin)	Chemotherapy (dactinomycin, cyclophosphamide, bleomycin, vincristine, doxorubicin) + radiotherapy + surgery	Cesarean section	34 weeks	No evidence of disease 4 years of discontinuing treatment	Normal
6.	Loguidice et al 1986[14]			Left iliac bone (skeletal)	None	Chemotherapy (cytoxan + Adriamycin)	Radiotherapy	Cesarean section		Stable disease 17 weeks after delivery	Normal
7.	Dhillon et al 1993[15]			Case 1- left scapula (skeletal) Case 2- right humerus (skeletal)	Case 1- left parietal area of skull Case 2- none	-	Radiotherapy, Chemotherapy(cyclophosphamide+ vincristine +doxorubicin)	Vaginal delivery		Case 1 – died 4 months after delivery Case 2 – died 6 months after delivery	Both babies normal
8.	Merimsky et al 1999[16]	1st month of pregnancy	Progressive right sciatic pain	Right sacroiliac joint (skeletal)	None	Chemotherapy (doxorubicin, ifosfamide, mesna)	Chemotherapy (doxorubicin, ifisfamide, mesna, etoposide), radiotherapy, surgery	Cesarean section	35 weeks	No evidence of disease 24 months after treatment	Normal
9.	Nakajima et al 2004[17]	25 weeks		Left thigh	None	Chemotherapy (doxorubicin, ifosfamide)	Radiotherapy, surgery	Cesarean section	32 weeks	died 7 months after delivery	Normal

10.	Dubios et al 2008[18]			Case 1- right ishium (skeletal) Case 2- 11th rib(skeletal)	Case 1 -lung, left frontal lobe, anterior abdominal wall Lung	Chemotherapy (vincristine, Doxorubicin, cyclophosphamide)	Radiotherapy	Case 1- cesarean section Case 2 – vaginal delivery		Case 1 – recurrence, no further follow up Case 2- disease freee 13 years after first recurrence	Normal newborns in both cases
11.	Blight & Puls 1981[19]			Retroperitoneal abdominal mass (extraskeletal)	None	-	Radiotherapy, surgery and chemotherapy (vincristine, cyclophosphamide, doxorubicin)	Vaginal delivery		Died 8 months after treatment	Normal newborn
12.	Gennatas et al 1987[20]			Left thigh	None	chemotherapy (vincristine, cyclophosphamide, Adriamycin, DTIC, actinomycin D)	chemotherapy (vincristine, cyclophosphamide, Adriamycin, DTIC, actinomycin D)	Cesarean section		Complete remission	Normal newborn
13.	Adiar et al 2001[21]			Duodenum (extraskeletal)	None	-	surgery and chemotherapy	Medical termination of pregnancy		No evidence of disease 10 months after treatment	
14.	Verheecke et al 2013[22]	41 weeks	Pain right leg	Right proximal tibia (skeletal)	Lung, abdominal cavity, breast	-	Chemotherapy (3 week alternating treatment of vincristine+ Adriamycin+ iphosphamide (VAC) and etoposide iphosphamide)	vaginal delivery	41 weeks	Brain metastasis discovered after 14 cycles of therapy	Normal newborn
15.	Ateser et al[23]	24 weeks	Generalised abdominal pain, nausea, vomiting	PNET ovary (extraskeletal)	Multiple	Surgery, Chemotherapy (doxorubicin, cyclophosphamide, vincristine)	chemotherapy	Cesearean section	37 weeks	Died due to progressive disease 13 months after the initial diagnosis	Normal newborn
16.	S. Schur et al[24]	18 weeks	Mass in right proximal lower limb	Pelvis with involvement of uterus (extraskeletal)	Lung	Chemotherapy (etoposide, ifosfamide, doxorubicin, vincristine)	Chemotherapy, surgery	Vaginal delivery	24 weeks	Disease free till 1 year after delivery	Intrauterine fetal demise after 2nd cycle of chemotherapy
17.	Al Khawaja et al[25]	16 weeks	Right sided back pain and gluteal pain	Right paraspinal mass in lumbar region with extension to first right sacral foramen and central canal (extraskeletal)	None	Surgical excision, chemotherapy (doxorubicin, cyclophosphamide, vincristine)	Radiotherapy, Chemotherapy (doxorubicin, cyclophosphamide, vincristine)	Vaginal delivery	32 weeks	Disease free till 9 months follow up	Normal baby

4. Conclusion

EFST in pregnancy is rare. Chemotherapy should be instituted early in the course of the disease and has shown favorable outcome in the past. Mode of delivery in these cases depends on the obstetric indication and site of the tumor as per the imaging findings. Incidence of malignancies in pregnancy though rare, is increasing due to delayed childbearing, and the clinicians should remain vigilant regarding complaints of the patient. The need of a through general physical examination cannot be overemphasized.

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6. Conflict of Interest

None.

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