EWING'S SARCOMA OR PRIMITIVE NEUROECTODERMAL TUMOUR OF CERVIX: A CASE REPORT

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HOW TO CITE THIS ARTICLE:
Sonali Deshpande, Kanan Yelikar."Ewing's Sarcoma or Primitive Neuroectodermal Tumor of Cervix: A Case Report". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 01, January 06; Page: 173-176.

ABSTRACT: Primitive neuroectodermal tumor (PNET) is extremely rare in the female genital tract but it has high grade malignant potential. EFT of cervix can be easily confused with other small round cell neoplasm. It reinforces the value of IHC in its objective identification especially at unusual sites where they are found to have the same aggressive behavior as those arising from usual sites & are similarly found to be responsive to optimum local treatment & combination chemotherapy with radiotherapy. We herein report the case of PNET presenting in intrapartum period.
KEYWORDS: Ewing's sarcoma, primitive neuroectodermal tumor of cervix, Immunohistochemistry.

INTRODUCTION: Although traditionally classified as separate entities; Ewing's sarcoma & peripheral primitive neuroectodermal tumor (pPNET) are now regarded as belonging to spectrum of neoplasm's exhibiting neuroectodermal differentiation& collectively referred to as Ewing's family of tumors (EFTs) having the same t(11;22) chromosomal translocation in >90% cases[1]. EFT of cervix is extremely rare & only 10 cases are described till date in English literature[2]. Here we present a case of EFT of cervix which stresses the importance of use of immunohistochemistry (IHC) in differential diagnosis of "small round cell tumors" occurring at unusual site.

CASE REPORT: A 27 year old G4P3L3 delivered a full term healthy child at zero hours of admission vaginally. Placental delivery was followed immediately by expulsion of a fleshy mass of size 5x3x3 cm. Clinical examination confirmed a similar friable, pedunculated, reddish-brown mass of size 8x6x6 cm, attached to the anterior lip of cervix with bilaterally free parametria. She was pale and rest of her clinical parameters were within normal limits. Provisional diagnosis as carcinoma of cervix stage $Ib_2$was kept.

She was unbooked & was apparently asymptomatic during antenatal period. On investigation her hemoglobin was 5.9g%, PCV 21.1%. Contrast CT scan was s/o 8x7x7 cm mass lesion involving cervix protruding into the vagina with mild contrast enhancement with maintained fat planes with surrounding structures with no intraabdominal thoracic pathology. Histopathology of expelled mass revealed encapsulated lesion composed of uniform cells with no mitosis suggestive of benign endometrial stromal nodule.

Patient underwent total abdominal hysterectomy with bilateral salphingo-oophorectomy owing to rapid regrowth of tumor, continuous blood loss attributable to the tumor, to determine the margins of the tumor required for diagnosis & to differentiate it from invasive stromal sarcoma after building up her hemoglobin levels.

On laparotomy uterus, bilateral ovaries, fallopian tubes & rest intra abdominal organs appeared normal, clinical findings confirmed. Histopathology of specimen revealed an infiltrating neoplasm with small to medium sized cells with occasional atypical mitotic figures suggestive of? EFT with normal endometrium, bilateral ovaries & fallopian tubes. Subsequent

immunohistochemistry (IHC) was confirmatory of EFT with very characteristic &sensitive CD99 positivity[2] with positivity for vimetin & CD56 with proliferative index Ki (60-70%). Cytogenetic studies could not be done due to lack of facility.

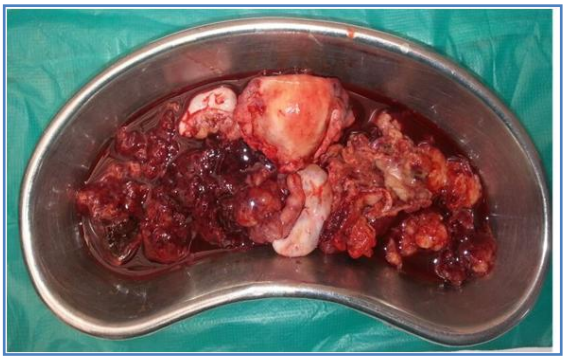
Patient didn't follow up & reported 3 months after surgery after pursuing her telephonically. Contrast CT scan was s/o 3.9x3x 4.8 cm mildly enhancing soft tissue lesion posterior to bladder, with multiple omental, bowel serosal& bilateral lung metastasis. She was started on "EFT protocol"- combination chemotherapy- vincristine (1.5mg/m^2), ifosfamide (2g/m^2), etoposide (100mg/m^2), cyclophosphamide (600mg/m^2), actinomycin (1mg/m^2)& radiotherapy. Patient is currently receiving third cycle of chemotherapy.

DISCUSSION: Primary extra osseous Ewing's sarcoma, also called primitive neuroectodermal tumor. PNET is a relatively uncommon tumor that mainly occurs in the trunk, limbs, and retro peritoneum[3].Primary cervical PNETs are extremely rare, and only 10 cases are reported in literature[2]. In contrast to patients with PNETs at other sites, which usually include children and young adults, the age of patients with primary cervical PNETs ranged from 21 to 50 years (mean, 34 years) [4].The patient presented in this report was 27 year old, which is consistent with literature. Lysyj andBergquist[5] reported the first case of Ewing's sarcoma in pregnancy in 1963.Their patient presented at the 32nd week of gestation with pain in the right leg and was diagnosed with Ewing's sarcoma of the pubic ramus and patient was delivered by caesarean section at 36 weeks of gestation. Neither chemotherapy nor radiation therapy was administered during gestation[5].Our patient was reported in intrapartum period and delivered a healthy baby vaginally. Farah Farzaneh et al[2] reporteda multiparousIranian women who presented with yellow purulent vaginal discharge since 3 months and on bimanual pelvic examination under general anesthesia revealed a 4× 5 cm mass apparently arising from the anterior lip of the cervix, producing yellow vaginal discharge, the size of uterus was around 10 weeks pregnancy. There was no extension of the lesion into vagina, parametria, or adjacent organs including the bladder and rectum. The tumor was clinically Ib2. The diagnosis of Ewing's sarcoma was supported by a strong membrane staining with CD99 which was very characteristic and sensitive. Our patient was relatively asymptomatic who delivered at zero hours of admission and placental delivery was followed immediately by expulsion of a fleshy mass of size 5x3x3 cm. Clinical examination confirmed a similar friable, pedunculated, reddish-brown mass of size 8x6x6 cm, attached to the anterior lip of cervix with bilaterally free parametria. She was pale & rest of her clinical parameters were within normal limits. Provisional diagnosis as carcinoma of cervix stage $Ib_2$was kept. We also did Subsequent immunohistochemistry (IHC) for confirmation of EFT with CD99 positivitywith positivity for vimetin & CD56 with proliferative index Ki (60-70%).

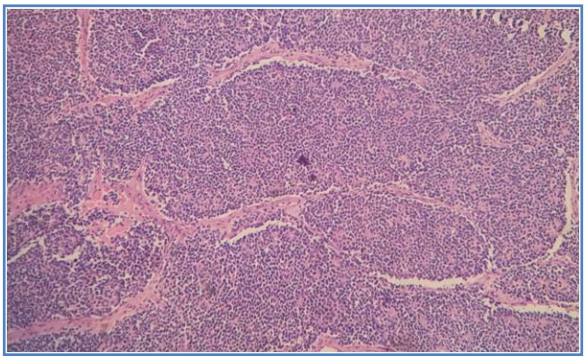
Ewing's sarcoma must be considered as a systemic disease without adequate treatment in which more than 90% of patients die from secondary haematogenous metastasis, occurring mainly in the lungs. The 5 year survival rate can increase to 55% to 60% with dose-intensive cytotoxic treatment regimens in localized disease; the 3 year disease-free survival rate was reported to be 15-22% among patients with detectable metastasis at the time of diagnosis[6].Our patient was also lost to follow up and reported after 3 months with multiple metastases. It might be considered that neoadjuvant chemotherapy is an overtreatment of PNET. However, as Snijders-Keilholz et al[7] mention, upto now duration of follow –up of the reported cases is too short (5-42 months) to prove or reject this discussion. Farah Farzaneh et al [2]reported that for Ewing's sarcoma of other sites,

neoadjuvant chemotherapy in metastatic cases could change inoperable presentation to operable state for successful local/or regional treatment. Our patient also reported with metastasis and now on chemotherapy.

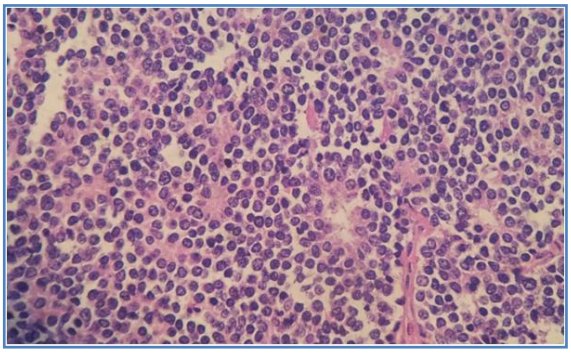
CONCLUSION: As in our case EFT of cervix can easily be confused with other small round cell neoplasms. It reinforces the value of IHC in their objective identification especially at unusual sites where they are found to have the same aggressive behavior as those arising from usual sites &are similarly found to be responsive to optimum local treatment & combination chemotherapy with radiotherapy. Presence of metastasis is the most important adverse prognostic factor.



Uterus with bilateral ovaries with tumour



Low power field picture



High power field picture

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Date of Submission: 11/10/2013.
Date of Peer Review: 12/10/2013.
Date of Acceptance: 17/12/2013.
Date of Publishing: 03/01/2014