Global Journals LaTeX JournalKaleidoscope™
Artificial Intelligence formulated this projection for compatibility purposes from the original article published at Global Journals. However, this technology is currently in beta. *Therefore, kindly ignore odd layouts, missed formulae, text, tables, or figures.*

Ewing Sarcoma of Ovary-An Unusual Presentation

Mohammad Faheem[1]

[1] Nuclear Medicine, Oncology and Radiotherapy Institute, Islamabad

Received: 12 December 2014 Accepted: 4 January 2015 Published: 15 January 2015

Abstract

Ewing?s sarcoma generally arises from bones and soft tissues. Primitive neuroectodermal tumor (PNET) and Ewing sarcoma constitute Ewing family of tumors. In International literature there have been rare reports of Ewing?s sarcoma as either primary tumor of ovary or as metastatic disease involving ovary. No such case has yet been reported in Pakistan. Here a case is being presented who was diagnosed as Ewing?s sarcoma of ovary. Workup showed no involvement of the bones.

Index terms— ewing?s sarcoma, PNET, ovary.

1 I. Introduction

wing's sarcoma is malignant round blue cell tumor which primarily arises from bones involving pelvis, femur, tibia, humerus and clavicle. Patients affected are commonly in their second decade of life. The definitive diagnosis is based on histomorphology, immunohistochemistry and molecular pathology. The pathologic differential diagnosis is the grouping of small round cell tumors which include lymphoma, alveolar rhabdomyosarcoma and others. Ewing's sarcoma typically has clear cytoplasm on H & E staining due to glycogen. Positive PAS staining proves the presence of glycogen. The characteristic immunostain is CD99 which diffusely marks cell membrane. The morphologic and immunohistochemical characteristics are corroborated with chromosomal translocations of which t(11;22)(q24;q12) is the commonest present in about 90% of Ewing' sarcomas 1 . Ovary is very rare site to give origin to Ewing's sarcoma. Few cases of ovarian Ewing's sarcoma have been reported in literature mainly affecting females 18-30 years of age and in all these cases diagnosis was made with the help of immunohistochemistry. Cases of Ewing's sarcoma of uterus, vagina and vulva have also been reported 2 . We are reporting a case of 30year old lady who was diagnosed as having Ewing's sarcoma of ovary.

2 II. Case Report

A 30 year old, premenopausal lady having 4 children with last child birth 4 years back and no significant family history presented at NORI Islamabad on 19 th February 2014. She had already undergone exploratory laparotomy on 12 th December 2013. She presented to gynecologist with complaint of pain lower abdomen. Her Abdominal USG was done on 8 th December 2013 which showed a large solid mobile mass in mid pelvis. The mass was not showing any relationship with major abdominal/pelvic organs. This was then followed by CT scan which showed a large well defined, smooth walled, complex soft tissue attenuation mass measuring 7.7x9.1 cm. The exact site of origin was difficult to ascertain on CT scan but it was probably in mesenteric fat. The lesion was mostly solid and differentials could be tumor of neural origin, lymphoma, GIST or carcinoid mass. There was additional finding of a suspicious mass in pelvis which was reported as either an adnexal mass or a nodal deposit (Fig A **??**nd B). This was then followed by exploratory laparotomy. CT scan report was not clear regarding the origin of pelvic mass. There were two masses peroperatively. One of the masses was measuring 4x4 cm and was related to posterior surface of uterus. The other mass measured 8x8 cm and was attached to fallopian tube. Excision of masses was done and histopathology showed Granulosa cell tumor of ovarian origin. Slide review was advised due to unusual presentation that was not in accordance with Granulosa cell tumor. Histopathology reviewed showed Ewing Sarcoma.

Immunohistochemistry demonstrated positivity of CD99 and NSE thus endorsing the diagnosis of Ewing Sarcoma. Postoperative CT scan was done that showed minimal pelvic ascites. She was planned for chemotherapy but our patient was then lost to follow up. She presented again to our hospital in July 2014 with complaint of pain abdomen. On examination there was a huge mass palpable in lower abdomen lower limit of which was not reachable. On pervaginal examination a mass could be felt outside vagina in relation to its posterior wall. CT scan was repeated that showed a large lobulated mesenteric mass and a pelvic mass with rectal wall thickness (Fig C **??**nd D). The case was discussed in MDM and it was decided to give her chemotherapy followed by evaluation for surgery. Vincristine, Doxorubicin and Cyclophosphamide were started alternating with Ifosfamide and Etoposide. Interval assessment was done. Although there was reduction in size of mesenteric and pelvic mass,(Fig E **??**nd F) however, surgery was still not possible due to indistinct fat planes with rectum. Chemotherapy was continued till 17 cycles. Doxorubicin was replaced with Dactinomycin after 5 cycles of Doxorubicin. Recent CT scan showed further reduction in size of pelvic mass and resolution of mesenteric mass. Fat planes with rectum became distinct. Currently, she has been referred for surgery after discussion in MDM.

3 III. Discussion

Formerly thought to be dissimilar Ewing sarcoma and PNET are now considered to be same tumors that demonstrate variable degree of neuroectodermal differentiation. Ewing sarcoma lacks neuroectodermal differentiation whereas PNET expresses neuroectodermal differentiation when evaluated either by microscopy or IHC 3 . Extraosseous Ewing Sarcoma is a rare entity and that of female genital tract is extremely uncommon. The commonest site of PNET in female genital tract is ovary followed by uterus. The paucity of the disease can result in diagnostic dilemmas 4 . Our patient was also initially misdiagnosed as a case of Granulosa cell tumor of ovary. In her case not only histopathology was incorrect but also imaging studies were misleading. The site of origin and accurate diagnosis was determined only after review of histopathology and immunohistochemistry. Had the patient been discussed in multidisciplinary meeting prior to embarking on surgery, in view of extensive disease she could have been advised neoadjuvant chemotherapy after guided biopsy. Another appropriate approach might have been diagnostic laparoscopy. Ovarian PNETs are very aggressive tumors and are associated with extremely poor prognosis due to high incidence of metastatic disease. Median survival ranges from 10.8 months to 3 years. The prognosis of patients presenting with localized tumors has improved in recent years by means of multimodality treatment such as surgery, radiation and chemotherapy. Chemotherapeutic agents frequently used are vincristine, doxorubicin and cyclophosphamide alternating with Ifosfamide and doxorubicin 5 . Although optimal debulking was done in this particular case as evident by postoperative CT scan but she didn't come for adjuvant treatment resulting in local recurrence within 4-5 months of surgery. In a case report by Anfinan et al local recurrence in 31 years old female was seen during adjuvant chemotherapy with vincristine, doxorubicin and Ifosfamide alternating with vincristine, adriamycin and Ifosfamide. Cases of Ewing sarcoma already reported in literature were in 18 to 30 years age group 2 and our patient was also 30 years old. This shows that patients to be affected with ovarian Ewing sarcoma are relatively young. A number of chromosomal abnormalities are associated with PNET/Ewing sarcoma including deletion of Retinoblastoma gene, ras homologue member I and overexpression of N-myc, fas ligand, tumor necrosis factor and epidermal growth factor receptor. These factors may perhaps be responsible for aggressive nature of these tumors 6 . Due to non availability in Pakistan chromosomal abnormalities were not studiedin this patient but CD99 and NSE were strongly positive on Immunohistochemistry. Our patient has responded well to chemotherapy, whether this translates into better survival warrants further follow up.

4 IV. Conclusion

Figure 1: Fig

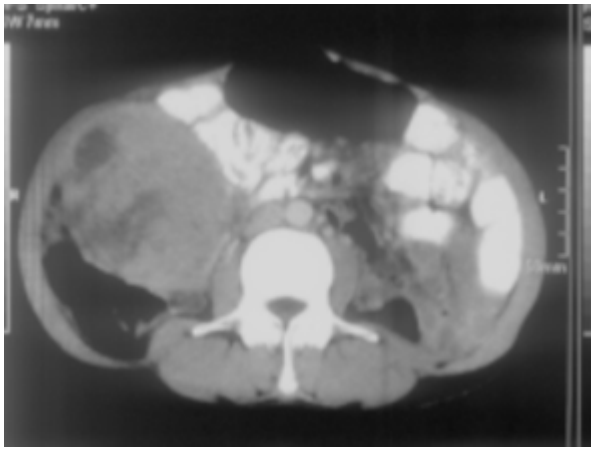

Figure 2: Fig C

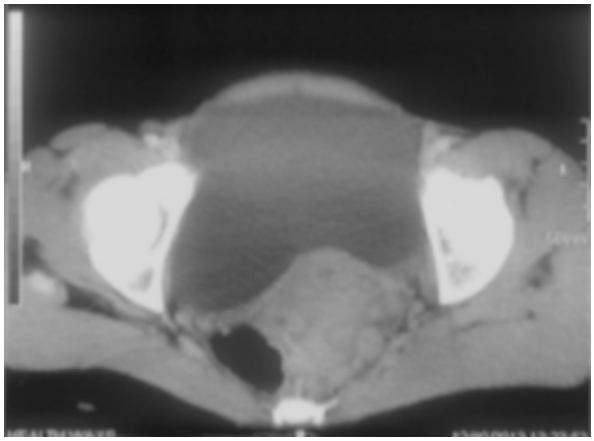

Figure 3: Fig E

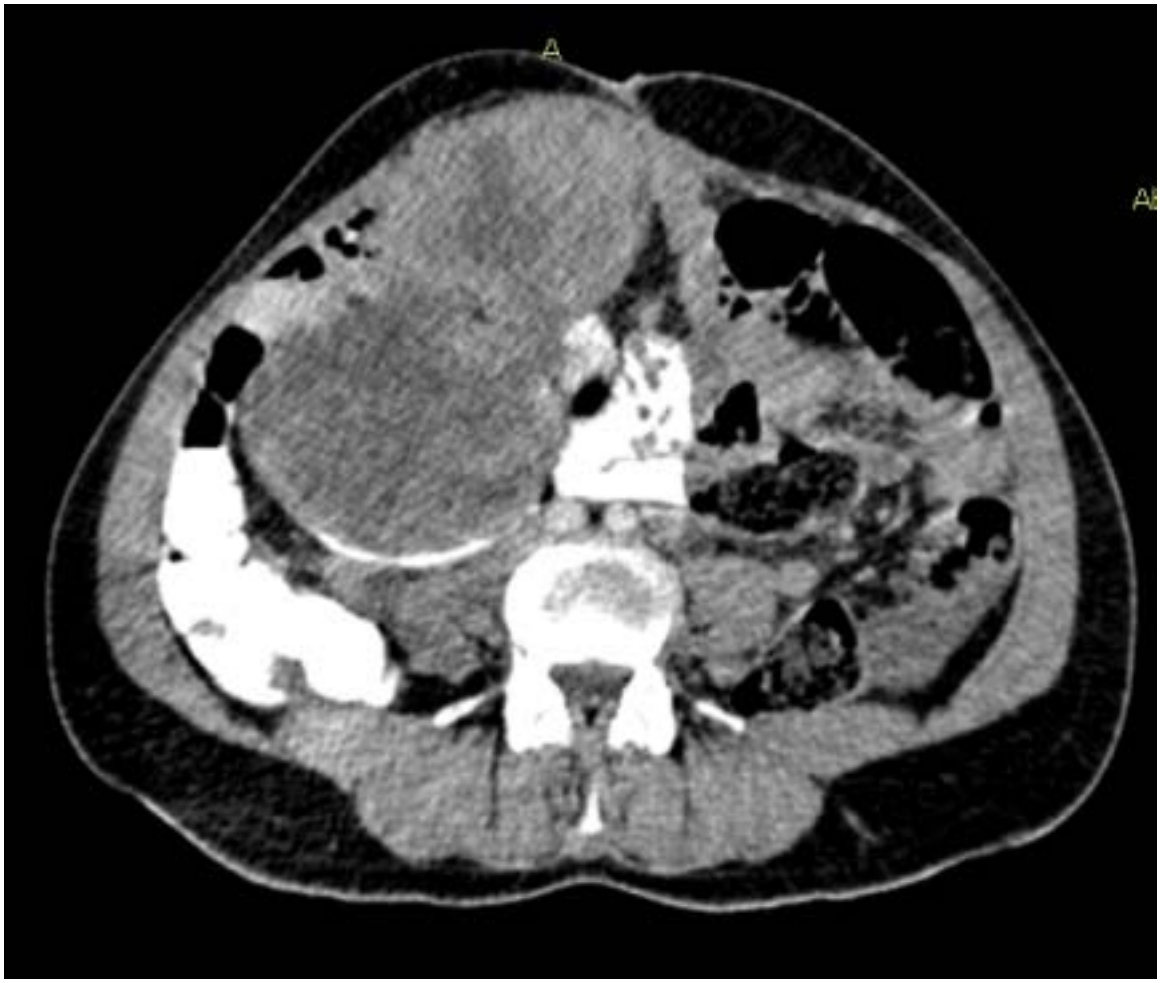

Figure 4: Fig G

[Kuk et al. ()] 'A case of primitive neuroectodermal tumor of the ovary'. J Y Kuk , S Y Yoon , M J Kim , J W Lee , B G Kim , D S Bae . *Korean J Obstet Gynecol* 2012. 55 (10) p. .

[Giuseppe et al. ()] 'Conservative treatment of Ewing's sarcoma of uterus in young women'. L Giuseppe , R Leonardo , D N Edoardo , Anna Mc Salvatore , A M Mario , V . 10.1155/2015/871821. http://dx.doi.org/10.1155/2015/871821 *Case Rep Obstet Gynecol* 2015. 2015.

[Mashriqi et al. ()] 'Ewing's sarcoma of cervix a diagnostic dilemma: a case report and review of literature'. N Mashriqi , J K Gujjarlapudi , J Sidhy , M Zur , M Yalamanchili . 10.1186/s13256-015-0733-2. *J Med Case Rep* 2015. 9 (1) p. 225.

[Cm et al. ()] 'Primary vaginal extraosseous ewing sarcoma/ primitive neuroectodermal tumor with cranial metastasis'. Yip Cm , Hsu , Ss , N J Chang , J S Wang , W C Liao , J Y Chen . *J Chin Med Assoc* 2009. 72 (6) p. .

[Anfinan and Sait ()] 'Primitive neuroectodermal tumor the ovary'. N M Anfinan , K H Sait , Al-Maghrabi , JA . *Saudi Med J* 2008. 29 (3) p. .

[Richard et al. ()] 'Sarcomas of Bone'. J Richard , G D Steven , Ahk Daphne . *Cancer Principles & Practice of Oncology*, V T Devita, T S Lawrence, S A Rosenberg (ed.) 2015. Wolters Kluwer Lippincott: Williams and Wilkins. p. . (10 th edition)