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Pediatric Ovarian Adenocarcinoma: Original Case Report.

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Abstract*: The Surface epithelial stromal tumours (adenocarcinomas), representing 80% to 90% of adult ovarian neoplasms, account for only 7% of malignancies in children . The objective of this study is to document a rare case of pediatric ovarian adenocarcinoma and to assess the efficacy of chemotherapy for treatment of such malignancies. A 14 years old female presented with abdominal distension and pain in abdomen.USG abdomen and pelvis revealed a complex ovarian cyst with normal CA-125 report. Patient undergone exploratory laprotomy with excision of ovarian mass. Histopathological report reviewed twice reported Mucinous cystadenocarcinoma. Post-operative USG was normal . Patient received 6 cycles of chemotherapy inj. paclitaxel 250 mg & inj Carboplatin 420mg from 30-8-15 to 20-12-15. Patient tolerated chemotherapy well. PET-CT Scan done on 29/03/2016 demonstrated ill defined FDG non avid hypodense lesion in right adnexa. Long –term follow up and disease free survival in under assessment.*

Keywords*: Chemotherapy, Mucinious Cystadenocarcinoma, pediatric Ovarian Adenocarcinoma, PET-CT Scan.*

I. Introduction

Ovarian malignancies in children may represent an array of unique problems for the clinician who is more accustomed to diagnosing and treating ovarian neoplasm in adults. Although ovarian malignancies in children are rare (representing 0.2% of all ovarian neoplasms), their recognition & diagnosis are vital because they can be fulminant if treated inadequately. Advances in combination chemotherapy have enabled patients to survive the disease with tolerable short term and long term morbidity. Whenever possible, treatment should be individualized to preserve reproductive and menstrual function without jeopardizing the life of the child. Surface epithelial tumours are rare in children and represent the same morbidity & mortality in children as in adult, therapy also is similar to that used in adults.

The average overall incidence of ovarian neoplasms is 1.7:100,000 per year [1,2]. The rarity of ovarian tumors in children precludes a statistically significant compilation of the age related occurrence of specific tumors. The surface epithelial stromal tumour, representing 80-90% of adult ovarian neoplasms[3], account for only 7% of malignancies in children [4]. Endometrial and clear cell carcinoma are not found in the first two decades of life [5,6]. The mucinous and serous cystadenocarcinomas are rare before puberty and have not been reported in children younger than 4 years old [7]. The incidence of these coelomic epithelial lesiosn increases with advancing age. The gross and microscopic characteristics of these tumours in adolescents are identical to those of similar neoplasms in adult. The tumors are bilateral in 10% of cases compared with 25% in adults.

The variety of symptoms associated with an enlarging ovarian lesion in pediatric patients commonly obscures the cause of the complaint and unduly delays diagnosis and treatment. Initial symptoms may be absent or vague and the average delay between the clinical onset and histologic diagnosis is 3 to 4 months[8]. The spectrum of symptoms referable to ovarian neoplasms in children is extensive. The most common presenting symptom is abdominal pain which is present in more than half of patients. Patients can also present with abdominal swelling with associated anorexia, weight loss, nausea or vomiting. The rarity of pediatric ovarian malignancies contributes to a low index of suspicion. Only 36% to 63% of cases are indentified correctly before surgery. A palpable ovary in the prepubertal patient is presumed to be abnormal because of the absence of gonadotropins. Although surgical resection and histologic examinations are the only definitive means of diagnosing an ovarian neoplasm, several radiographic, ultrasonographic , and laboratory parameters can aid the clinician.

Although malignant potential of surface epithelial stromal tumours in children (7.1 % to 13.5 % of cystadenomas) is less than is seen in adults, malignant neoplasm in children show a clinical course and mortality similar to what is seen in adults . Serous lesions have the poorest prognosis. Tumours of low malignant potential behave in a generally benign fashion, as if properly staged as IA, may be treated with salpingo- oopherectomy only. For malignant surface epithelial stromal tumors other than stage IA treatment includes total abdominal hysterectomy, bilateral salpingo- oopherectomy, omentectomy, pelvic / para-aortic lymph node sampling. Chemotherapy generally indicated for adjunctive therapy [1,9,10]. Radiotherapy with a

moving strip technique may be beneficial. The 10 year survival rate for all stages is 75 % primarily because of the preponderance of early stage lesions [9].

II. Case

A 14 years old female presented with pain in abdomen and abdominal distension. Clinically, large mass of 32-34 weeks was palpable per abdomen with smooth surface and regular margins. USG abdomen and pelvis done on 19/04/2015 was suggestive of complex ovarian cyst of size 12 x 17cm with multiple loculi and septations. Serum CA 125 was 50 U/ml. patient undergone exploratory laprotomy with excision of ovarian mass on 20/04/2015. Histopathology report was suggestive of mucinous cystadenocarcinoma. Slides and blocks were reviewed as age is uncommon for ovarian epithelial malignancy. USG abdomen and pelvis on 04/07/2015 showed post operative status and rest was normal. Chemotherapy for 6 cycles in interval of 3 weeks was given from 30/08/2015 to 20/12/2015 (inj paclitaxel 250 mg and inj carboplatin 420 mg). patient tolerated chemotherapy well. CT scan abdomen and pelvis done on 09/01/2016 revealed right ovary bulky 4x2.6x3.8 cm with heterogenous parenchymal enhancement and left ovary not visualized post operative status. CTscan chest revealed multiple well defined tiny soft tissue lesions with adjacent interstitial nodular thickening in apical segment of right upper lobe. Soft tissue nodule in superior segment of right lower lobe (multiple lung metastasis with adjacent lymphatic spread). Serum CA125, Serum AFP, Serum beta HCG and serum LDH were all within normal limits.Patient was asymptomatic for lung metastasis. PET-CT scan was done on 29/03/2016 which showed an ill defined hypodense lesion in right adnexa and was FDG non avid. No evidence of FDG avid pelvic or retroperitoneal lymphadenopathy. Tiny FDG non avid lung nodule was there on PET-CT which is of uncertain significance and needs follow up. No other FDG avid distant metastasis was there. Now patient is kept on close follow up. Long term disease free survival is under assessment.

III. Figures

Fig. 1

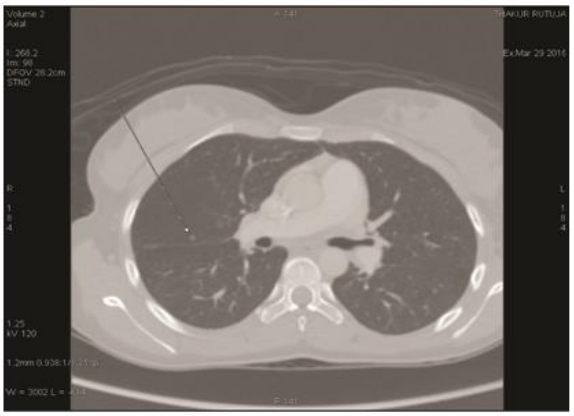


Fig. 2

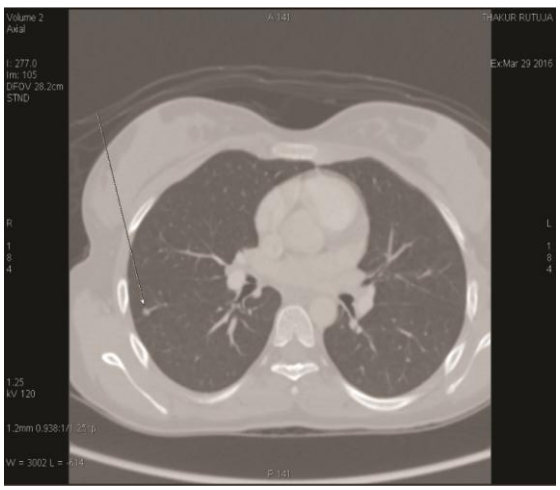


Fig. 3

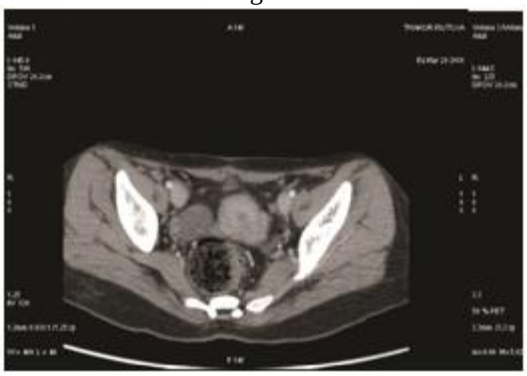
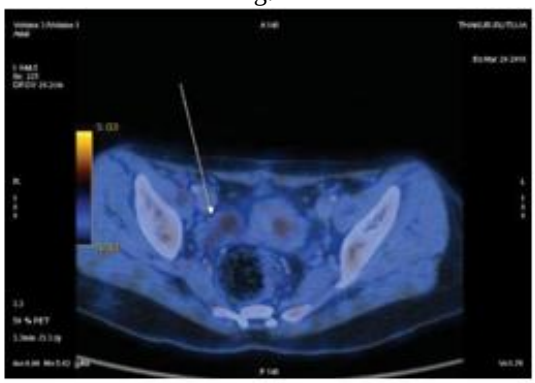


Fig. 4



PET CT SCAN images showing tiny FDG non-Avid lung nodules and ill defined FDG non Avid hypodense lesion in right adnexa.

IV. Discussion

Pediatric ovarian Adenocarcinoma is a rare entity. Serum CA-125 is used as a prognostic indicator as in adult ovarian malignancies. Surgical line of treatment and chemotherapy regimens are same in pediatric ovarian Adenocarcinoma as in adults. Review of 67 cases of pediatric ovarian tumors was done by Kris Ann P. Shultz et al in which 30 patients had benign tumours, 37 patients had malignant tumours; 11 immature teratomas, 7 malignant mixed germ cell tumours, 7 juvenile granulose cell tumours,5 dysgerminomas,2 endodermal sinus tumours,2 serous papillary cystadenocarcinomas,1 small cell carcinoma,1 anaplastic sex cord tumor and 1 undifferentiated sarcoma. PET-CT scan should be considered as a follow up tool for disease and metastasis assessment. The aim of this study is to have an emphasis over such rare malignancies, so that proper treatment protocols could be followed and disease can be cured at an early stage.

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