Complex cystic abdominal mass in an adolescent girl

Introduction

Ovarian tumors are rare in children with a reported incidence of 2.6 cases per 100000 population per year. Cystic masses in abdomen are commonly seen in children and the differential diagnosis include ovarian cysts, cystic mesenchymal hamartoma of liver, duplication cysts of the alimentary tract, omental cysts, retrotroperitoneal cystic teratoma and large lymphangiomatous lesions. In addition there are many organ specific cysts seen with a lesser frequency. When the cysts are too large to be assessed by an US scan- first imaging modality currently used- diagnostic errors are common. If the child has presented with respiratory embarrassment, leading to orthopnea, a diagnostic or therapeutic paracentesis may be required, if there is doubt as to the organ of origin, especially in the absence of facility for an urgent MRI scan. Once the diagnosis is confirmed as an ovarian cyst, all attempts are made to perform a fertility preserving surgery; the recommended surgery is a Unilateral Salpingo Oophorectomy in cases of a Borderline Tumor of the Ovary. The following case report exemplifies the difficulties in initial assessment and management.

Case report

BA, a 12 years old girl was admitted through the ER, for inability to lie flat on the bed for approximately two weeks associated with some reduction in appetite in the preceding month. She had no bowel or urinary symptoms; and her LMP was on the day of presentation; with menarche 11months earlier. She denied any history of jaundice or bleeding disorder in the past. She weighed 39.7 KG, which was in the 3rd centile for her age. On examination she was not anemic and had no significant peripheral lymphadenopathy. Her abdomen was massively distended and one could not discern any organomegaly. The abdomen appeared fluid filled and was dull on percussion with minimal dilated veins on the abdominal wall. Investigations showed normal white cell count and ESR. The Serum AFP, Beta HCG, LDH and Liver function tests and renal function tests were normal. As the mass was not highly suspected to be ovarian in origin, a Ca 125 level or CEA level were not done at this stage. Next an US scan was done. This showed a giant fluid filled mass filling almost the entire abdomen, measuring over 38cms in the longitudinal axis and over 24cms in the transverse axis. The "cyst" was extending from pelvis to xiphi-sternum, compressing the bladder was seen partially filled. As an MRI scan was not immediately available, a CT scan with contrast was done (Figure 1 & 2).

This showed a giant fluid filled mass, non-enhancing and without any calcification. The mass had faint septa; but did not show multi-locaulation. The mass was compressing the liver and the diaphragm upwards and the bowel loops to the left upper quadrant of the abdomen. One could discern a normal ovary on the left side; but none on the right side. The radiologist felt that a fluid filled mass of lymphangiomatous origin was the first of the possibilities; with ovarian pathology as the next. This was in view of multiple septa seen on US scan, and normal biochemistry and normal tumor markers. As the fluid filled mass was causing orthopnea, it was decided to do a percutaneously drainage using a small tube, at the part where the mass was closely adherent to the abdominal wall, in the low right iliac fossa, using a 6F pigtail catheter. The procedure was uneventful and over the next 3 days, about 7800ml fluid drained. The fluid was almost colorless and cytological examination of the fluid done showed NO malignant cells or atypical cells. The weight of the patient reduced by 8.6KG, with complete relief of orthopnea. She was able to have full normal diet and normal bowel actions. An ultrasound scan was repeated and this confirmed that there was no free fluid in the peritoneal cavity. The cyst was still very large, and the computed residual fluid volume in the mass was 664 ml.

from abdominal cavity with multiple abdominal packs first. Next a double layer of purse string sutures were placed around the tube to avoid any fluid spill into the wound or abdominal cavity. A right salpingo-oophorectomy was done, after confirming that the opposite side ovary and tube were completely normal (Figure 3).

The liver, omentum and retro peritoneum were all normal. A thorough peritoneal lavage was done before closure, taking care to send the lavage returns for cytology- that was later reported as normal, with no malignant cells. Patient made an uneventful recovery and was discharged on the 3rd day. We received the final histology as follows. The cyst shows features of a Mucinous Cyst adenoma with Borderline Malignancy. No evidence invasion of the wall and no tumor of the external surface. Tubes showed evidence of salpingitis isthmica nodosa. FIGO classification Stage 1A (Figure 4).

Discussion

Ovarian neoplasms are rare in pediatric population forming only 2.6/100000 cases per year1 In a series of 1037 malignant tumors published from Texas,2 the age adjusted incidence of ovarian malignancy was only 0.102 and 1.072 in children 0-9years and 10-19years per 100000 per year, respectively. Thus, ovarian

DOI: 10.15406/jpnc.2019.09.00385
malignancy is rare in children and adolescence, but they are the commonest genital tumors accounting for almost 60-70% of gynecological malignancies in this age group. Ovarian tumors comprise of both benign and malignant varieties, both solid and cystic types. Germ Cell Tumors, surface epithelial stromal tumors and miscellaneous tumors like lymphoma, leukemia and small cell carcinoma. WHO has published a classification of ovarian tumors as Low grade Epithelial Carcinoma, with IV Taxane/Carboplatin-3-6 cycles. In our patient the fluid cytology, as well as the peritoneal washouts were negative for any malignant cells and there was no evidence of any malignant cell on the surface of the mass making it a FIGO Stage 1A.

Summary

A 12 year old girl presented with massive abdominal distention causing orthopnea. Despite ultrasonography and CT scan examination, diagnostic difficulties of its organ of origin and the need to give symptomatic relief, necessitated controlled paracentesis of the fluid filled mass. A borderline ovarian tumor of 8.6 KG weight was removed intact, by a fertility preserving right salpingo-oophorectomy, sparing the opposite normal ovary and tube and uterus. Final histology confirmed a Stage 1A Borderline Ovarian Tumor of the Mucinous Cystadenoma type. The case is reported for its rarity in this age group and is the first one reported in this country.

Acknowledgments

The authors gratefully acknowledge the Director General of Royal Hospital for permission to publish patient related data for academic purposes. We would like to acknowledge and thank Dr Fatima Ramadhan Al Lawatia, Senior Consultant, Department of Pathology for the slides and the legend for the slides. Dr Thuria Al Rawahi, Senior Consultant, Department of Gynecology gave us suggestions regarding oncological aspects of the case and her help is gratefully acknowledged. Dr Mohammed Jaffer Sajwani as head of department of Pediatric surgery gave us help and encouragement for publishing this report. No grants or financial assistance was received in publishing this case report.

Conflicts of interest

The authors declared there is no conflict of interest.

References
