Ovary-preserving tumorectomy for immature teratoma in an adolescent – Case report

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Summary

The authors present a case of a 14-year-old premenarchal girl with a large solid tumor of the left ovary. The rim of normal ovarian tissue was visible around the tumor on ultrasonography scan. Although the levels of two tumor markers, LDH and CA125, were elevated, the authors performed an organ-sparing tumorectomy. The final pathology report revealed foci of immature neural tissue, with a final diagnosis immature teratoma Stage Ia.

Key words: Ovary; Tumor; Ultrasound; Immature teratoma.

Introduction

The ultimate goal in the treatment of pediatric and adolescent patients with ovarian tumors is to preserve the tube and the ovary whenever possible. Normal ovarian tissue or ovarian crescent sign relies on the fact that healthy ovarian tissue can be seen adjacent to the cyst or tumor within the ipsilateral ovary [1]. This morphological ultrasound sign has a potential to become a simple and effective way of excluding an invasive ovarian malignancy [1, 2]. The crucial dilemma that arises when deciding the extent of surgery in adolescent with large solid ovarian tumor is whether to perform ovariectomy or adnexectomy when ovarian crescent sign is present, or to go for ovary-sparing tumorectomy.

Case Report

A 14-year-old premenarchal girl was referred for abdominal discomfort. She was otherwise healthy with negative family history for breast/ovarian disease or malignancy. Abdominal palpation revealed a mobile mass extending from the pelvis to the umbilicus. Abdominal ultrasound examination showed a 16-cm, predominantly solid tumor, arising from the left ovary. A rim of normal ovarian tissue was observed adjacent to the tumor, within the ovary (Figures 1A and 1B). Doppler pulsatile index was 1.3 in peripheral vessels. To eliminate bias the patient was assessed independently by two gynecologists with sonographic experience longer than 15 years with ovarian tumors. The serum level of LDH was 1054 IJ/l and of CA 125 - 216 mIJ/l, while alpha-fetoprotein, beta hCG, inhibin B, CA 19-9, estradiol, testosterone, neuron specific enolase, and lab works - electrolytes, liver function tests, and urine analysis, were within normal limits. The laparoscopy was excluded due to the tumor volume and structure, and presence of the ovarian crescent sign lead the authors to consider tumorectomy as a treatment of choice. Upon entering the abdominal cavity, a large smooth mass was found arising from the left ovary. There were few proliferative changes in the omentum and peritoneal surface in the pelvis. Peritoneal washings were obtained. A superficial linear incision was made along the antimesenteric border of the ovarian tumor, approximately three cm from the ipsilateral tube, parallel to it. Next, the thinned ovarian cortex was gently peeled off the tumor wall by blunt and sharp dissection (Figure 1C). The goal was to enucleate the neoplasm without opening it (Figures 2A and 2B). The tumor weighed 1,000 g. It was sent to frozen section analysis, which revealed benign mature teratoma with preponderance of mature neural elements. The authors reconstructed the remaining ovarian tissue. Appendectomy was performed due to enlargement of the appendix, and due to the presence of neural elements; they proceeded with surgical staging, including biopsy of peritoneum, omentum, and pelvic lymph nodes. The blood loss during surgery was 300 ml. The patient was discharged from the hospital on the fifth day. Histopathology reported a mostly mature teratoma with few foci of immature neural tissue within the tumor mass. The lymph nodes, peritoneal washing, and appendix showed no presence of tumor tissue. Biopsy of the omentum revealed peritoneal deposits of mature glial tissue -gliomatosis peritonei. The final report was immature teratoma of the left ovary, Stage Ia, Grade I. Six-week follow up serum levels of LDH and CA 125 were normal, and the volume of the affected ovary was 7.5 cm^3 (Figure 3). At 18 months follow up menarche occurred and ultrasonography showed no signs of relapse.

Discussion

Laparoscopic cystectomy is a safe and effective method of managing ovarian dermoid cysts in the pediatric and adolescent patient population [3]. In a low percentage of dermoid foci of immature teratoma can be found. The dilemma arises when deciding the type of surgery in adolescents with great solid ovarian teratoma. The ovarian crescent sign as a morphological ultrasonographic feature was first described in adult and later in pediatric and adolescent patients with ovarian masses [1, 2]. The authors reported that in children and adolescents who were surgically treated for benign adnexal

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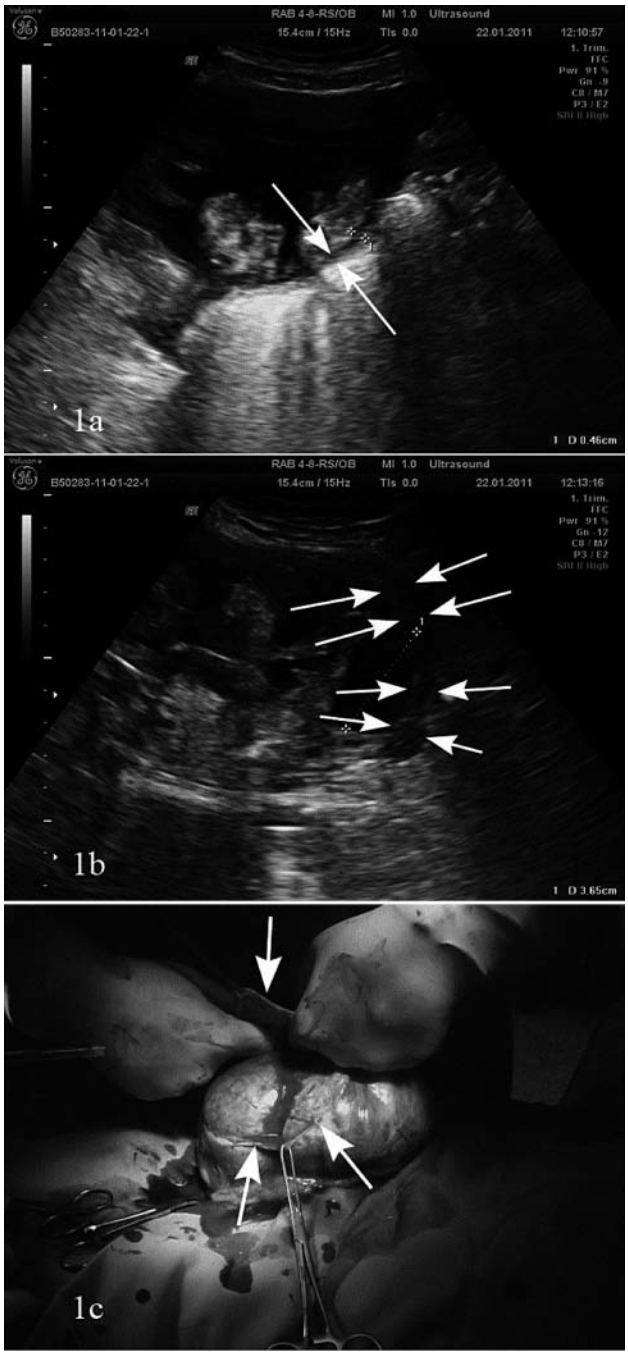

Figure 1A – Ultrasonographic appearance of a rim of normal ovarian tissue (arrows).
Figure 1B – Functional cyst between a rim of normal ovarian tissue and tumor (arrows).
Figure 1C – Appearance of an ovarian cortex peeled off the tumor (arrows).

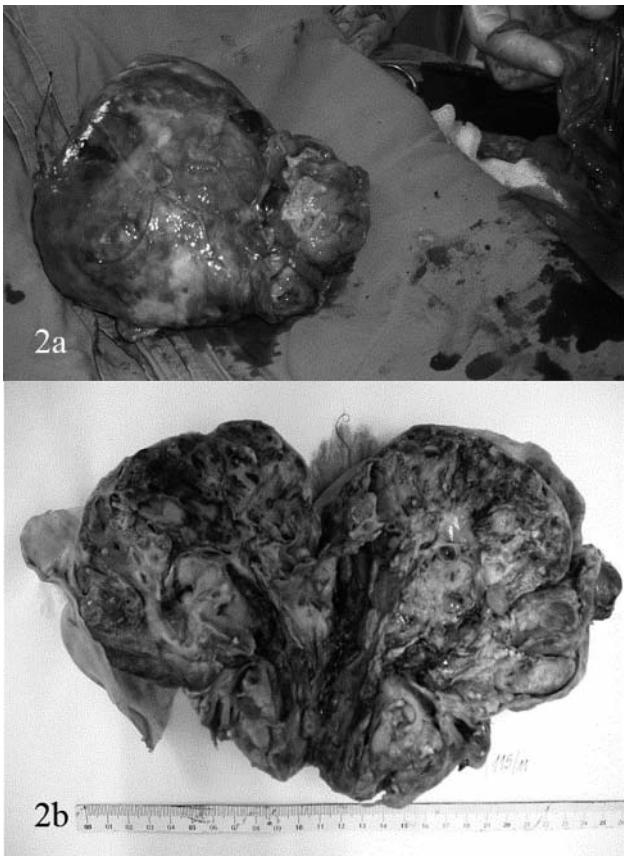

Figure 2A – Extirpated tumor and preserved whole ovary in the hand of surgeon.
Figure 2B – Macroscopic view of the tumor.

Figure 3 – Ultrasonographic appearances of both ovaries six weeks after surgery.

masses, the absence of the ovarian crescent sign was found in one-third of the masses complicated with torsion [2]. Absence of the ovarian crescent sign does not exclude a benign tumor, however its presence lowers the probability of an invasive ovarian malignancy in both adult and pediatric and adolescent patients [1, 2]. The absence of the ovarian crescent sign should be an indicator to refer a patient to a gynecologist specialized in practice with pediatric and adolescent population [2]. The pulsatility index appeared to be less sensitive than the ovarian crescent sign and the morphology index in dis-

criminating between malignant and benign tumors, which is similar to the reports on adult patients [2, 4]. Serum levels of LDH may be elevated in mature teratomas. Adding a single CA 125 measurement to the ultrasound imaging performed by an experienced examiner does not improve preoperative discrimination between benign and malignant adnexal masses even in adult patients [5]. It is generally accepted that small foci of immature neural elements can be readily missed on frozen section, especially if the tumor is large. Only very large immature teratomas Grade I, Stage 1 (greater than 1,500 g) warranted consideration of adjuvant chemotherapy [6]. Peritoneal gliomatosis occurs in 10% of patients with immature teratomas, and is biologically of benign nature [7]. An adolescent with immature teratoma Stage I Grade I was treated by Einarsson *et al.* with laparotomy and adnexectomy without chemotherapy. A full surgical staging procedure was based on preponderance of mature neural elements in the tumor on frozen section [8]. The authors decided against chemotherapy, based on the tumor weight and the stage of the disease. Six weeks similar as 24 months after surgery, they measured ovarian volume, which showed no significant difference between affected and contralateral ovary; without evidence of damage of ovarian reserve that was reported for laparoscopic excision of ovarian cysts [9].

Conclusions

The authors reported that the presence of the ovarian crescent sign may be a useful ultrasonographic morphological feature, which could support preservation of ipsilateral ovary, even in early stage of malignancy in adolescents. The visualization of healthy ovarian tissue does not require a high level of ultrasound skills and could be successfully included into a routine ultrasonographic practice of all operators who find adnexal tumors in young patients. Further prospective work could show whether the use of the ovarian crescent sign may provide a more effective path of managing pediatric and adolescent patients with adnexal masses.

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