Pseudo-Meigs syndrome secondary to endodermal sinus tumor

Dilan Altintas Ural,1 Ali Erdal Karakaya,1 Ahmet Gokhan Guler,1 Can Acipayam,2 Mustafa Sabih Kaya,2 Mehmet Cihan Karacaoglu,3 Sezen Kocarslan4

1Department of Pediatric Surgery, Kahramanmaraş Sutcu Imam University Faculty of Medicine, Kahramanmaraş, Turkey
2Department of Child Health and Diseases, Kahramanmaraş Sutcu Imam University Faculty of Medicine, Kahramanmaraş, Turkey
3Department of Radiology, Kahramanmaraş Sutcu Imam University Faculty of Medicine, Kahramanmaraş, Turkey
4Department of Pathology, Kahramanmaraş Sutcu Imam University Faculty of Medicine, Kahramanmaraş, Turkey

ABSTRACT

Ovarian tumors are the most common gynecological tumors seen in girls. Approximately 60–70% of them are germ cell tumors. Pseudo-Meigs syndrome is characterized by the presence of pelvic tumoral mass (benign or malign), pleural effusion and massive acid. If the tumor is removed, acid and hydrothorax disappear. Endodermal sinus (yolk sac) tumor is a very rare cause in the diagnosis of pseudo-Meigs syndrome, and only a few cases have been reported. This case is one of the rare cases presenting with pseudo-Meigs syndrome and pathologically diagnosed as yolk sac tumor.

Keywords: Acid; pleural effusion; pseudo-Meigs Syndrome; yolk sac tumor.

CASE REPORT

A 12-year-old girl was brought with abdominal pain, which started about 10 days ago, with a complaint of abdominal mass. The patient had significant abdominal distention on physical examination (Fig. 1). In diagnosis, abdominal ultrasonography (USG) was performed first. Abdominal computed tomography (CT) and abdominal magnetic resonance imaging (MRI) were performed as advanced imaging methods. On chest X-ray, an effusion was detected in the right hemithorax.

Abdominal USG determined a 17X10 cm vascular solid lesion in the pelvis and widespread fluid in the pelvis at a depth of 11 cm. In contrast-enhanced whole abdominal CT, a cystic necrotic lesion with a solid com-
ponent starting from the right part of the pelvis and con-
trasting multiple irregularly at a size of 17x13x9 cm, and
10 cm free acid in the pelvis was detected (Fig. 2A). MR
detected a massive mass lesion of 15x12x9 cm extending
in the inferior bladder and uterus in the abdomen, 10
cm free fluid in the pelvis (Fig. 2B). The patient's tumor
markers were as follows: beta subunit of human chori-
onic gonadotropin ($\beta$-HCG): 4.3 mlu/ml (2–6), alpha
fetoprotein (AFP): 26900.0 IU/ml (0–5.8).

Informed consent was obtained from the patient’s
parents. Following approval of informed consent and
operation preparations, the patient underwent surgery.
Approximately 4.5 liters of free acid was aspirated in
the operation. The mass adhering to the liver originat-
ing from the right ovary and adhering to the omentum
from the upper part with a smooth contour was removed
by right salpingo-oophorectomy (Fig. 3). Samples were
taken from peritoneum and omentum. The pathological
diagnose was reported as a yolk sac tumor with a size of

15x13x8 cm (Fig. 4). Omentum and peritoneal tissues
were biopsied. In fluid histopathology, mesothelial cells
and rare lymphocytes were seen. The patient was given
Bleomycin- Etoposide- Cisplatin (BEP) chemotherapy
protocol by pediatric oncology.
DISCUSSION

Ovarian tumors are the most common gynecological tumors in children and is approximately 1% of all childhood cancers. 40–50% of the ovarian masses are neoplastic, and 60–70% of these are germ cell tumors. Epithelial tumors are seen rarely (10–15%). Germ cell tumors develop from embryonic gonad primitive germ cells. These tumors include dysgerminoma, yolk sac tumor, embryonal carcinoma, mature teratoma, immature teratoma, choriocarcinoma and malignant mixed germ cell tumors. All germ cell tumors are malignant except mature teratoma [5].

Endodermal sinus tumor (yolk sac tumor) is a very rare cause in the diagnosis of pseudo-Meigs syndrome, and only a few cases have been reported [6, 7]. The cause of the acid has not been cleared, it is thought to be due to the transudation than its absorption capacity towards the tumor surface. Another reason is the formation of a peritonitis due to the growth of the tumor [8].

The most common symptoms of ovarian tumors are abdominal pain and mass. Other findings are urinary accumulation, constipation, anorexia, vomiting and intestinal obstruction due to the compression effect of the mass. It has aggressive growth and spreading potential. Tumor size can be up to 30 cm in diameter, with an average diameter of 15.5 cm. In most patients, symptoms appear in less than a week. If complications such as torsion, cyst rupture and perforation occur, the patient suffers from acute abdominal syndrome [5].

USG gives information about the size, structure and localization of the ovary. Advanced imaging methods (CT and MRI) are recommended to assess pelvic anatomy, paraaortic lymph nodes, and detect the presence of metastases [9].

Tumor markers are important in postoperative follow-up and regression of the disease. AFP, beta-HCG, CA-125, LDH, CEA and CA-19-9 levels are less specific in children than adults for the ovarian malignant tumors. In contrast, beta HCG and AFP are more sensitive in children. Yolk sac tumor of ovary releases AFP. In our case, the level of AFP was quite high (AFP: 26900.0 IU/ml (0–5.8).

Yolk sac tumor may be associated with other germ cell tumors. Microscopically the most common subtype is the reticular type which has a structure named Schiller-Wall body. This structure consists of primitive cells around a capillary. The treatment requires surgery. In fertility-sparing surgery, unilateral oophorectomy, salpingo-oophorectomy and intraabdominal tumor debulking are the possible techniques to remove the gross tumors. Bleomycin- Etoposide- Cisplatin (BEP) and Vinccristine-Actinomycin D- Cyclophosphamide (VAC) are commonly used chemotherapy regimens [10].

In conclusion, although rarely seen in the causes of Pseudo-Meigs syndrome in girls, yolk sac tumor should also be considered in differential diagnosis. Fertility-sparing surgery should be planned as the most appropriate treatment and patients should be followed up for long-term.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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