

Ovarian Mature Cystic Teratoma Containing Homunculus : A Case Report

We report a partial fetus-like structure (homunculus) in benign ovarian mature cystic teratoma removed from a 23-yr-old female. The cyst displayed various tissues derived from the three germ layers. The homunculus had a distinguished fetal cranial structure with one upper extremity. A partially developed osteocartilagenous skeleton was identified in the cranial structure of the homunculus. Intracranial structures such as cerebral cortex, retinal pigment, and leptomeninges were extruded from the partially disrupted calvarium.

Key Words : Ovarian Neoplasms; Dermoid Cyst; Homunculus; Teratoma

**Yong Ho Lee, Sung Gun Kim,
Sung Hyuk Choi*, In Sun Kim†,
Sun Haeng Kim**

Department of Obstetrics and Gynecology, Department of Emergency Medicine*, Department of Pathology†, College of Medicine, Korea University Hospital, Seoul, Korea

Received : 31 October 2002
Accepted : 20 December 2002

Address for correspondence

Yong Ho Lee, M.D.
Department of Obstetrics and Gynecology, Korea University Hospital, 126-1 Anam-dong 5ga, Sungbuk-gu, Seoul 136-705, Korea
Tel : +82.2-920-5837, Fax : +82.2-921-5357
E-mail : leejuu@lycos.co.kr

INTRODUCTION

The Latin word "homunculus" literally means "a structure resembling a miniature human body" to designate a human being not produced by pregnancy and this definition is used in morphologic entities. This tumor should be distinguished from the more highly developed fetus-in-fetu, a malformed parasitic monozygotic twin that is found inside the body of the living child or adult.

We report a case of a mature cystic teratoma containing a fetus-like structure (homunculus).

CASE REPORT

A 23-yr-old woman was admitted to the hospital with a history of palpable abdominal mass of 4 weeks' duration. The patient had never experienced pregnancy, and her menstrual periods were regular. The medical history and review of systems were unremarkable. Physical examination revealed a huge pelvic mass of soft consistency and free mobility. Abdominal ultrasound demonstrated bilateral cystic masses containing a solid portion in both ovaries. Routine laboratory data taken at admission were normal.

At exploration, bilateral huge ovarian cystic masses, measuring about 15 cm in diameter with a smooth glistening surface in both, were identified. The uterus and both fallopian tubes were normal. Bilateral cystectomy was performed

for preservation of future fertility and the removed tissue was sent for pathologic examination. Enzyme immunoassay for urinary human chorionic gonadotropin was negative immediately after the operation. The postoperative course was uneventful, and the patient was discharged on the fourth postoperative day.

Gross Findings

The cyst of the left ovary had a gray membranous surface and measured about 15 cm in diameter. On opening the mass, there was another cyst, 5 cm in diameter. The smaller mass was filled with hair intermixed with greasy yellow sebaceous material and yellowish clear fluid.

The right ovarian dermoid cyst had a reddish-brown, smooth, and glistening external surface and measured 15 cm in diameter. It was traversed by many small tortuous blood vessels and was composed of multilocular cysts in continuity. In the largest one, there was a small mass similar to a human body of head and single upper extremity, measuring about 5 cm in its greatest dimension (Fig. 1). The epidermal surface of homunculus was uniformly rough and covered by hair. Osteocartilagenous structure surrounded by fat was noted after dissection. Skeletal muscles, ligaments, parenchymal organs, or organoid structures were not identified grossly within the homunculus. The upper portion of the homunculus was reminiscent of primitive appearance of head, and the distal portion had one appendage exhibited single upper extremity. On the

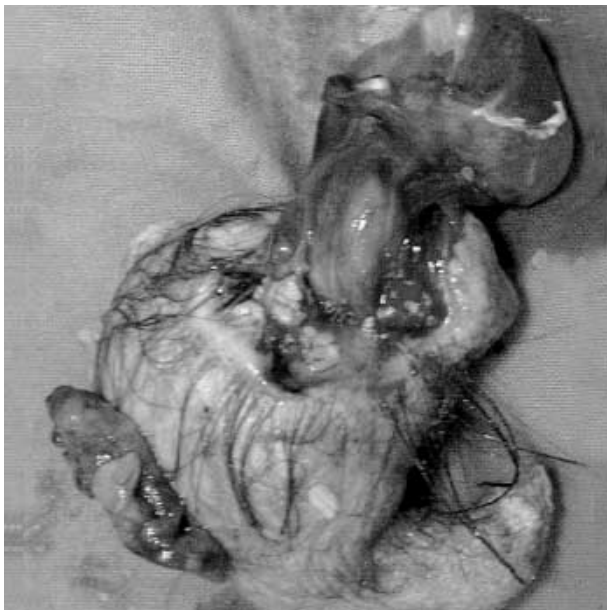


Fig. 1. Posterior view of the homunculus showing the herniated brain tissue through partially disrupted calvarium.

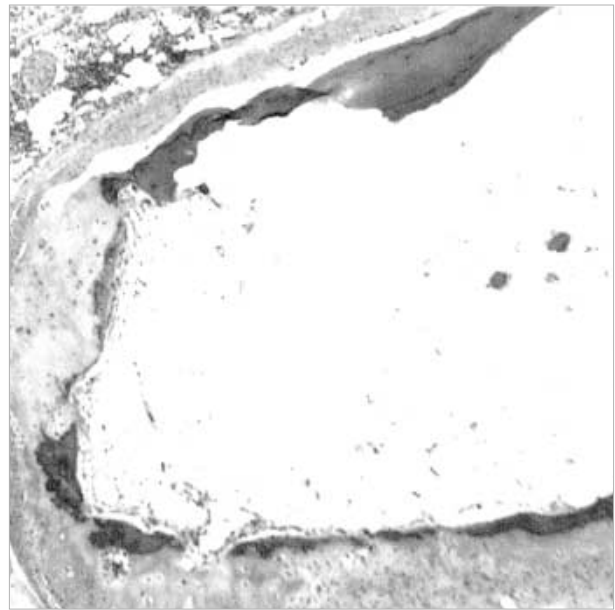


Fig. 2. Section from the caudal portion shows the tubular bone composed of central fatty marrow and peripheral bone and cartilage plate (H-E, ×80).

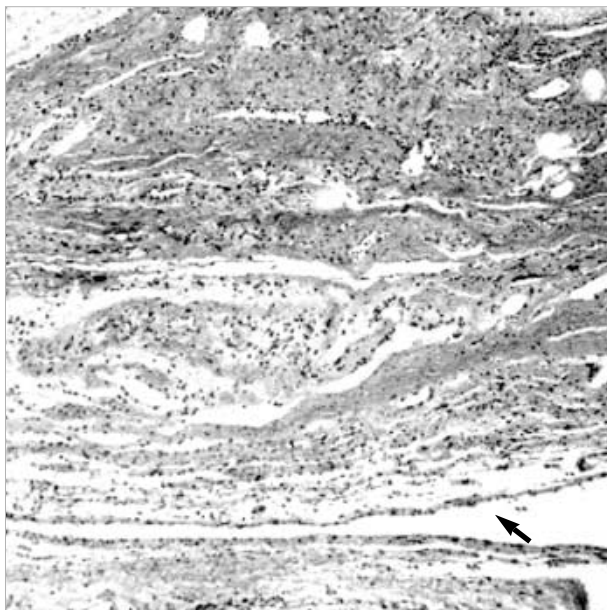


Fig. 3. Section from the cranial portion with a cyst discloses brain tissue covered by leptomenigeal lining (arrows) (H-E, ×80).

posterior aspect of the upper portion, sac like structure was protruding from the bony defect of the cranium lined by hair.

Microscopic Findings

The right ovarian tumor was benign cystic teratoma lined by normal skin and with attached adnexal components such

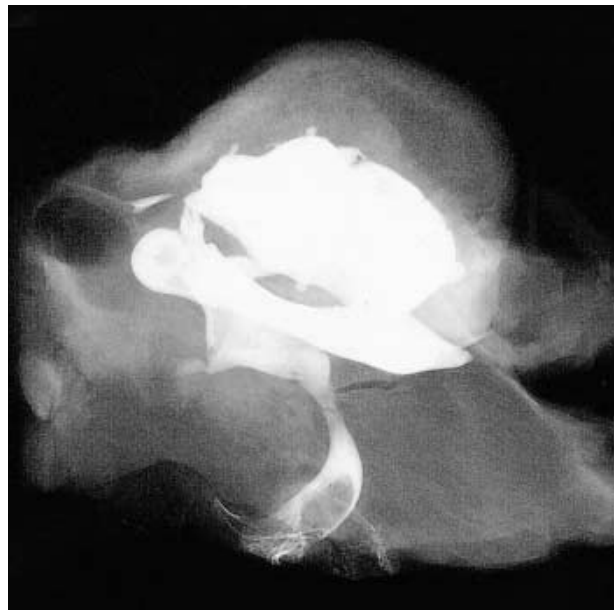


Fig. 4. Roentgenographic findings of the cranial portion of the homunculus revealing temporal bone, zygomatic arch, and mandible.

as primitive or mature hair follicles, sebaceous glands, and eccrine glands. Mature adipose tissue constituted the bulk of the tumor, and minor seromucinous gland components were seen. Tubular bones in the distal portion were composed of central fatty marrow and outer shell of lamellated bone with cartilage plate on the ends (Fig. 2). The caput-like structure in the upper portion was partly covered by skin and partly by

leptomeninges with retinal pigments, and composed of convoluted cerebral cortical tissue (Fig. 3). The flat bone in this area was also composed of central fatty marrow and peripheral rim of lamellated bone.

The left ovary was also cystic and lined by skin and its annexae. Foreign body reaction to ruptured epidermal components was seen.

Roentgenographic Findings

Radiographic examination of the homunculus revealed a calcific density in the superior portion (cranium) of the specimen (Fig. 4). It resembled the temporal bone, zygomatic arch, temporomandibular joint, and mandible. The caudal portion showed a tubular bony structure (Figure not shown).

DISCUSSION

Mature cystic teratomas, commonly called dermoid cysts, are the most common benign germ cell tumors of the ovary, and are encountered predominantly in women in their second and third decades of life. Histologically, they display a varying admixture of elements of one or more of the three cell lines: ectodermal, endodermal, and mesodermal tissue derivatives.

Despite the benign nature of these neoplasms, considerable interests have been paid on them because of the unusual histogenesis caused by the totipotency of these tumors that may give rise to any body structures or tissues and even the structure similar to a fetus.

Among the previously reported mature cystic teratomas or dermoid cysts of the ovary, only 24 cases of teratoma containing homunculus have been reported (1-6). In many cases, the caudal region of the homunculus, including the lower extremities, is typically more developed than the cephalic portion and some cases (5, 6) of prominent cephalic region had been reported as in our case. Despite a number of conflicting theories (7-10) to explain the histogenesis of teratomas, the most probable ones are misplaced blastomere and parthenogenetic development of a germ cell. Whatever the reason for the development of teratomas may be, these tumors seem-

ingly arise from the totipotential embryonic cells broken away from the normal developmental pathway to form the various fetal tissues.

This case represents the developmental pattern of the upper part of the human body rather than the lower part. It showed a head-like structure with a single upper extremity. The head-like structure contained temporal bone, zygomatic arch, temporomandibular joint, and mandible. The brain tissues, such as leptomeninges, retinal pigments, and cerebral cortex, were identified through the bony defect of the skull. The upper extremity representing the bony density of tubular structure had the central fatty marrow and the rim of lamellated bone. There were no internal organs as in the other reports.

In summary, we report here a rare case of homunculus in mature cystic teratoma of the ovary.

REFERENCES

1. Miyake J, Ireland K. *Ovarian mature teratoma with homunculus coexisting with an intrauterine pregnancy*. *Arch Pathol Lab Med* 1986; 110: 1192-4.
2. Inoue M, Mitsuda N, Tanaka Y, Suehara N, Ueda G, Kurachi K. *Benign cystic teratoma of ovary containing a homunculus*. *Nippon Sanka Fujinka Gakkai Zasshi* 1985; 37: 1050-3.
3. Abbott TM, Hermann WJ Jr, Scully RE. *Ovarian fetiform teratoma (homunculus) in a 9-year-old girl*. *Int J Gynecol Pathol* 1984; 2: 392-402.
4. Weldon-Linne CM, Rushovich AM. *Benign ovarian cystic teratomas with homunculi*. *Obstet Gynecol* 1983; 61: 88S-94S.
5. Azoury RS, Jubayli NW, Barakat BY. *Dermoid cyst of ovary containing fetus-like structure*. *Obstet Gynecol* 1973; 42: 887-91.
6. Plaut A. *Dermoid cyst of ovary containing a structure similar to a human body (homunculus)*. *J Mt Sinai Hosp* 1945-46; 12: 567.
7. Sherrer C, Gerson B, Woodruff JD. *The incidence and significance of polynuclear follicles*. *Am J Obstet Gynecol* 1977; 128: 6-12.
8. Ashley DJ. *Origin of teratomas*. *Cancer* 1973; 32: 390-4.
9. Bolande R. *Teratomas, Cellular Aspects of Developmental Pathology*. Philadelphia: Lea & Febiger: 1967.
10. Willis RA. *Pathology of Tumors*. 4th ed. New York: Appleton-Century-Crofts: 1967; 995.