

Multiple dermoid cysts within the same ovary: Our experience of a rare case with review of the literature

**N. Bournas¹, M.D.; M. Varras¹, M.D., Ph.D.; D. Kassanos¹, M.D., Ph.D.; Ch. Chrelias¹, M.D., Ph.D.;
O. Tzaida², M.D.; E. Salamalekis¹, M.D., Ph.D.**

¹Third Department of Obstetrics and Gynaecology, ²Department of Pathology, University General Hospital "ATTIKON", Athens,
Third District National Health System, Athens (Greece)

Summary

Objective: Ovarian dermoid cysts are relatively frequent ovarian lesions that can be easily diagnosed by transvaginal ultrasonography. However, the recognition of multiple dermoid cysts, separated by normal ovarian tissue, within a single ovary is rare.

Case: We present a case with bilateral dermoid cysts, four dermoid cysts within the right ovary and one in the contralateral ovary. Transvaginal ultrasonography showed hyperechogenic areas in the right ovary with a hypoechogenic area in front of them; the hyperechogenic areas were separated by areas with the echogenicity of normal ovarian tissue and indentation, suggesting the presence of multiple dermoid cysts within the same ovary.

Conclusion: Preoperative ultrasonographic suspicion of multiple dermoid cysts within the same ovary might contribute to a better decision about the surgical approach.

Key words: Dermoid cyst; Mature teratoma; Multiple; Same ovary; Unilateral; Ultrasonography.

Introduction

Dermoid cysts, commonly known as mature teratomas or benign cystic teratomas, are relatively frequent ovarian lesions, accounting for about 5-25% of all neoplasms of the ovary [1]. In approximately 10-15% of cases, the tumors present on both sides [2]. They usually occur frequently in young patients and microscopically one may find all types of ectodermal, mesodermal, and endodermal elements [3].

The clinical course of dermoid cysts of the ovary is commonly asymptomatic and they are usually discovered as a pelvic mass during physical examination or radiologic studies performed for other purposes. In cases of symptomatic dermoids, they can cause abdominal pain or swelling [1]. Due to continuous growth and possible complications including torsion of the ovary, rupture, infection and malignant degeneration, dermoid cysts should be surgically removed [2]. Uterine adnexal torsion or spontaneous rupture of ovarian dermoid cysts occurs in 16% and 3-7% of patients, respectively [4]. Malignant transformation of dermoid cyst of the ovary is rare, with an incidence of 1.8% [5].

Despite the frequency of dermoid cyst of the ovary, the appearance of multiple synchronous ovarian teratomas within the same ovary is a rare occurrence [6]. Moreover, the ultrasonographic appearance of this entity is not well described. We found only one case of an ultrasonographic description of multiple unilateral ovarian teratomas by Johnson and Jordan in 2001 [6]. The purpose of the present study is to describe the ultrasonographic findings of a case with multiple dermoid cysts within the same ovary and to discuss the differential diagnosis of this entity.

Case Report

A 23-year-old, gravida 0, para 0 woman, presented to the emergency department of a general hospital in Athens because of fever (39.6°C), right lower quadrant pain and diarrhea. She had a one-month history of abdominal discomfort after physical fatigue. Also, she complained of dyspareunia for the previous six months but she had no associated dysmenorrhea or body weight loss. Her menarche had been at age 12 years. Her menstrual periods were irregular (oligomenorrhoea up to six months); for this reason she was treated with oral contraceptives. During the previous year she had not used oral contraceptives and her menstrual cycles had been regular. A pelvic ultrasound scan was without any abnormal findings two years before and there was no history of diabetes or hypertension.

On admission, physical examination disclosed a well-nourished, hemodynamically stable woman in no acute distress. Her abdomen was soft and no peritoneal irritation signs were elicited. Laboratory studies revealed the following: hemoglobin, 11.9 g/dl; hematocrit, 39%; white blood cells, 8.700/μl; polymorphonuclear leukocytes 70.4%; platelets, 287,000/μl; normal values for blood urea nitrogen, serum electrolytes, prothrombin time 12.8 sec. and activated partial thromboplastin time 41.7 sec. The diagnosis of viral gastroenteritis was made and the patient was allowed to return home on symptomatic therapy, with the recommendation to see a gynecologist.

The next day the patient was examined at the Outpatient Gynecologic Clinic of the Third Department of Obstetrics and Gynaecology, University General Hospital "ATTIKON", Athens. Bimanual examination demonstrated a normal-sized uterus and a definite non-tender mass in the right adnexa. A questionable mass on the left was noted. Transabdominal sonography demonstrated a normal-sized uterus containing normal endometrial echoes. A cystic mass with echogenic elements was detected in the right ovary (Figure 1). Transvaginal ultrasonography showed the presence of hyperechogenic areas in the right ovary, with a hypoechogenic area in front of them (Figure 2a). The hyperechogenic areas were separated by

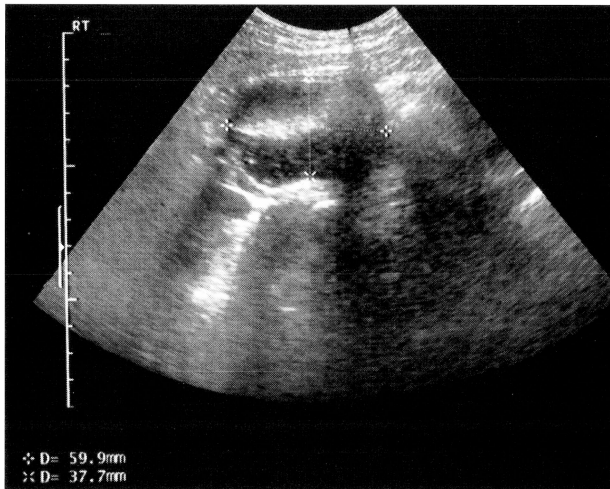


Figure 1. — Transabdominal ultrasonography of a part of the right ovary shows a cystic mass (6 x 3.8 cm) with echogenic elements.

normal ovarian tissue echoes (Figure 2b, arrow a) and indentations (Figure 2b, arrows a and b). All the hyperechogenic areas showed round contour. We suspected that there were probably more areas with dermoid cysts but it was difficult to separate them. At vaginal ultrasonography a solid left adnexal mass with diffuse hyperhechoic lines was also revealed; the maximum diameter was 3.5 cm. Laboratory tests for serum CA125, α -fetoprotein, β -human chorionic gonadotropin, carcinoembryonic antigen and CA19-9 were normal.

On exploratory laparotomy, through a Phannenstiell incision, two weeks following ultrasound examination both ovaries were found enlarged and cystic and covered by a thick white capsule. The right ovary contained four distinct cystic masses, separated by ovarian tissue; these masses were resected without any rupture of the walls. Then, deep interrupted sutures were placed to close the ovary; the ovarian surface was closed with the "baseball technique" [7]. Also, a 3.5-cm left ovarian cyst was enucleated with preservation of the ovary. With the operative findings and the ultrasonographic findings of the separate hyperechogenic areas we understood the situation (Figure 3).

The patient tolerated the procedure well and was discharged on postoperative day 5.

Grossly, the four cystic tumors from the right ovary were round with the maximum diameter ranging from 3 cm to 5 cm. Each of them had a smooth external surface and was fully surrounded by a rather thick glistening capsule with prominent vessels (Figure 5a). Their consistency was doughy and at sectioning they contained sebaceous material and matted hair. The largest one was bilocular with readily recognizable nodules (the dermal papilla) (Figure 5b). The cystic mass from the left ovary was macroscopically circumscribed with a smooth external surface and measured 3.5 x 3 x 2.5 cm. The cut surface was also filled with smegma and hairs (Figure 6).

Microscopic evaluation of each of the five cysts revealed the presence of mature adult-type tissues derived from two or more germ cell layers. (a) Ectodermal elements: epidermis with skin adnexal structures (hairs, follicles, sebaceous and sweat glands) were present in all of them, whereas neural tissue with the form of ganglion cells was recognized in only one. (b) Mesodermal elements: smooth muscle tissue, cartilage and adipose tissue were identified in some. (c) Endodermal elements of the type of bronchial epithelium and thyroid tissue were recognized in the largest right cystic tumor.

Discussion

Dermoid cysts are congenital tumors that contain derivatives of all three germ layers. They contain sebaceous material and hair, and their walls are lined in part by squamous epithelium, which contains hair follicles and sebaceous glands. Teeth, bone, cartilage, thyroid tissue and bronchial mucous membranes are often found in dermoid cysts. They have fatty components, with or without calcifications [8]. Recognition of multiple dermoid cysts, separated by normal ovarian tissue, within a single ovary is rare. In 1901, Winston reported the first case of two separate and distinct dermoid cysts of the right ovary [9]. Subsequently, in 1906, Kelly described two cystic teratomas in one ovary [10]. Later, in 1909, Novak described a patient with 21 dermoids, 11 in one ovary and ten in the contralateral gonad [11]. In 1925,

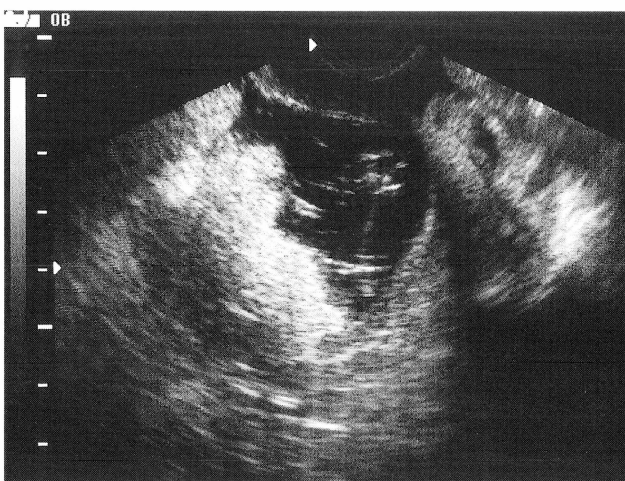


Fig. 2a

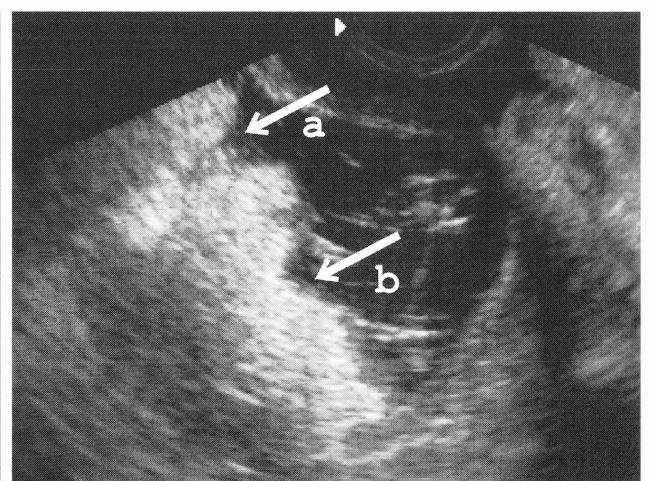


Fig. 2

Figures 2a & 2b — Transvaginal sagittal ultrasonography demonstrates the presence of hyperechogenic areas in the right ovary with a hypoechogenic area in front of them. Note the indentations between the hyperechogenic areas (arrows a, b). All the hyperechogenic areas show round contour.

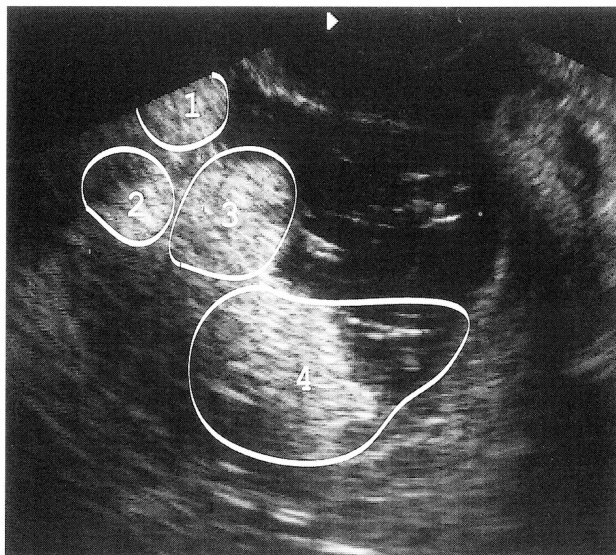


Figure 3. — Explanation of Figure 2 after the operative findings: separate hyperechoic areas represent multiple dermoid cysts (1, 2, 3, 4) in the right ovary.

Koncky reported seven cases of multiple ovarian dermoids in the same ovary among 100 cases of benign cystic teratomas [12]. In 1978, Imperato-McGinley identified multiple dermoid cysts throughout both ovaries of a patient with polycystic ovarian disease [13]. In 1982, Damewood *et al.* reported two cases of multiple dermoid cysts of the ovary; in the first case both ovaries contained four distinct dermoid cysts, while in the second case the left ovary contained three separate dermoid cysts [14]. More recently, Johnson and Jordan described a patient with bilateral dermoid cysts; one in the left ovary and four in the contralateral ovary [6].

Various theories regarding the origin of ovarian teratomas have been proposed, such as incomplete twinning,

autofertilization, blastomere inclusions, parthogenesis or disordered asexual growth of a primordial germ cell with variable embryonic differentiation [6, 14]. It is possible that a genetic defect may be responsible for the disruption of the first division of the meiotic cycle with resultant parthogenesis. The familial occurrence of teratomas, the presence of dermoid cysts in twins and multiple ovarian teratomas within the same ovary could be explained by a single gene mutation affecting the first division in meiosis [14]. Also, the presence of a 46,XX karyotype found in most ovarian dermoid cysts supports the theory of parthogenesis [15].

In recent years, transvaginal ultrasonography has dramatically improved the accuracy of ultrasonographic diagnoses, minimizing errors and allowing proper counseling and treatment [16]. Dermoid cysts have many and varied ultrasonographic appearances depending on their internal consistency. In general, the most common ultrasonographic appearance of a dermoid cyst is a complex, predominantly solid mass with echogenic internal components arising from fat and the calcified portion of the mass. Less common appearances of dermoid cysts range from the totally cystic dermoid cyst, encountered in those dermoid cysts lined by neuroectoderm, to the almost completely solid dermoid cysts that have a large amount of soft tissue component within. If the fat content is significant, they may be overlooked on ultrasound, as the echogenicity of the fat may be misinterpreted as adjacent bowel or surrounding tissues. Very characteristic findings of dermoid cysts are the presence of hyperechoic areas and acoustic shadowing from teeth or calcification within a semisolid semicystic mass [17, 18]. The distinction between hyperechoic solid appearing areas within a single dermoid cyst versus multiple dermoid cysts may be recognized with difficulty at ultrasonography. The intense posterior acoustic shadowing seen in many ovarian dermoid cysts may obscure the margins of the tumor and the ovary, preventing recognition of sepa-

Fig. 4a

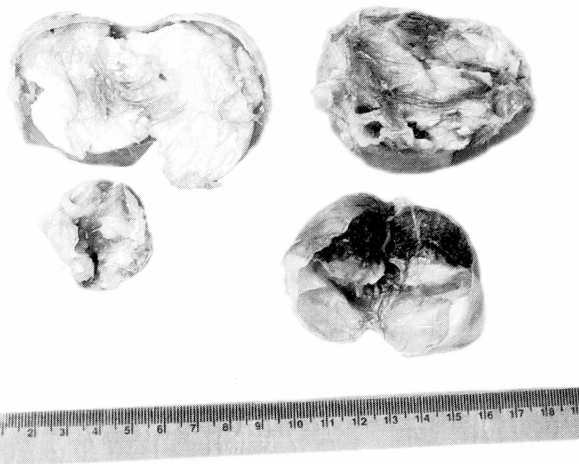


Fig. 4b

Figure 4. — **a)** Macroscopic appearance of the four cystic masses from the right ovary. The maximum diameters fluctuate from 3 cm to 5 cm (scale represents cm).

b) At section, the cysts contained smegma and hair. The largest cyst was bilocular; elements of cartilage and bone were identified. Dermoid cysts were confirmed by histopathology.



Figure 5. — Cut section of gross specimen showing smegma and hair within the dermoid cyst (3.5 x 3 x 2.5 cm) from the left ovary (scale represents cm).

rate masses [17, 18]. In our case, we suspected the presence of multiple hyperechogenic areas because of the two indentations between the hyperechogenic areas (Figure 2b, arrows a, b) and the space between two of these areas (Figure 2b, arrow a), although the intense posterior acoustic shadowing obscured the margins of the dermoid cysts and particularly the margins of dermoid cyst 2 in Figure 3.

The differential diagnosis of multiple dermoid cysts within a single ovary includes (i) coexistence of a solitary ovarian dermoid cyst with hemorrhagic cysts or endometriomas or multilocular cystic masses within the same ovary; (ii) coexistence of a solitary ovarian dermoid cyst with a primary ovarian neoplasm or metastases in the same ovary; (iii) coexistence of a solitary ovarian dermoid cyst with a teratoma arising from either the greater omentum or a supernumerary ovary; (iv) malignant transformation of a dermoid cyst with the malignant component simulating a second ovarian mass; (v) mucinous tumors arising in dermoid ovarian cysts; (vi) extra-ovarian adnexal disease that obscures the ovarian margins, such as pelvic inflammatory disease, endometriosis or inclusion cysts [19].

In conclusion, indentations between the hyperechogenic areas of ovarian dermoid cysts seen at vaginal ultrasonography as well as the existence of normal ovarian tissue between the cysts should increase the suspicion of a preoperative diagnosis of multiple dermoid cysts within the same ovary, therefore making a better decision as to the surgical approach.

References

- [1] Serafini G., Quadri P.G., Gandolfo N.G., Gandolfo N., Martinoli C., Derchi L.E.: "Sonographic features of incidentally detected, small, nonpalpable ovarian dermoids". *J. Clin. Ultrasound*, 1999, 27, 369.
- [2] Mecke H., Savvas V.: "Laparoscopic surgery of dermoid cysts - intraoperative spillage and complications". *Eur. J. Obstet. Gynecol. Reprod. Biol.*, 2001, 96, 80.
- [3] Disaia P.J., Creasman W.T.: "The adnexal mass and early ovarian cancer". In: Disaia P.J., Creasman W.T. (eds.). *Clinical Gynecologic Oncology* St Louis, Mosby-Year Book, Inc. 1997, 266.
- [4] Morgante G., Ditto A., Marca A., Trotta V., De Leo V.: "Surgical treatment of ovarian dermoid cysts". *Eur. J. Obstet. Gynecol. Reprod. Biol.*, 1998, 81, 47.
- [5] Ueda G., Fujita M., Ogawa H., Sawada M., Inoue M., Tanizawa O.: "Adenocarcinoma of the benign cystic teratoma of the ovary: report of a case with a long survival period". *Gynecol. Oncol.*, 1993, 48, 259.
- [6] Johnson S.C., Jordan G.L.: "Sonographic diagnosis of multiple unilateral ovarian teratomas". *J. Ultrasound Med.*, 2001, 20, 279.
- [7] Davis J.: "Adnexal surgery". In: Gershenson A.H., DeCherney A.H., Curry S.L. (eds.). *Operative Gynecology*. Philadelphia, W.B. Saunders Company, 1993, 513.
- [8] Rathod K., Kale H., Narlawar R., Hardikar J., Kulkarni V., Joseph J.: "Unusual "floating balls" appearance of an ovarian cystic teratoma: sonographic and CT findings". *J. Clin. Ultrasound*, 2001, 29, 41.
- [9] Winston D.Y.: "Two separate and distinct dermoid cysts and one serous cyst of the right ovary". *Tr. M. Soc. Tennessee*, 1901, 200.
- [10] Kelly H.A. *Operative Gynecology*, 1906, 2, 308.
- [11] Novak J.: "Über multiple dermoide der ovarien". *Beitr. Pathol. Anat. Pathol.*, 1909, 14, 1.
- [12] Konckly J.D.: "Ovarian dermoids: A study of one hundred consecutive cases". *Ann. Surg.*, 1925, 81, 821.
- [13] Imperato-McGinley J., Peterson R.E., Sturla E., Dawood Y., Bar R.S.: "Primary amenorrhea associated with hirsutism, acanthosis, nigricans, dermoid cysts of the ovaries and a new type of insulin resistance". *Am. J. Med.*, 1978, 65, 389.
- [14] Demewood M., Rosenshein N.B., Woodruff J.D.: "Multiple benign cystic teratomas of the ovary. Report of two cases and review of the literature". *Diagn. Gynecol. Obstet.*, 1982, 4, 243.
- [15] Inoue M., Fujita M., Azuma C., Saji F., Tanizawa O.: "Histogenetic analysis of ovarian germ cell tumors by DNA fingerprinting". *Cancer Res.*, 1992, 52, 6823.
- [16] Ferrari M.M., Mezzopane R., Bulfoni A., Grijuela B., Carminati R., Ferrazzi E., Pardi G.: "Surgical treatment of ovarian dermoid cysts: a comparison between laparoscopic and vaginal removal". *Eur. J. Obstet. Gynecol. Reprod. Biol.*, 2003, 109, 88.
- [17] Lindsell D.R.M.: "Pediatric Gynaecological Ultrasound". In: Bates J. (ed.). *Practical Gynaecological Ultrasound*. Greenwich Medical Media LTD, London, 1997, 161.
- [18] Fleischer A.C.: "Sonographic evaluation of adnexal masses with transabdominal or transvaginal sonography". In: Fleischer A.C., Javitt M.C., Jeffrey R.B. Jr., Jones H.W. (eds.). *Clinical Gynecologic Imaging*, Philadelphia, Lippincott-Raven Publishers, 1997, 48.
- [19] Ronnett B., Seidman J.: "Mucinous tumors arising in ovarian cystic teratomas: relationship to the clinical syndrome of pseudomyxoma peritonei". *Am. J. Surg. Pathol.*, 2003, 27, 650.

Address reprint requests to:
M.N. VARRAS, M.D., Ph.D.
Platonos 33
Politia (Kifisia)
14563 Athens (Greece)
E-mail: mnvarras@otenet.gr