CASE REPORTS

Two twins with teratoma of the ovary. An unusual association: case report

M. Indinnimeo[1], *M.D.*; C. Cicchini[1], *M.D.*; A. Larcinese[1], *M.D.*; S. Kanakaki[1], *M.D.*; F. Ricci[2], *M.D.*; P. L. Mingazzini[2], *M.D.*

[1]Department of Surgery "Pietro Valdoni", [1]Department of Experimental Medicine and Pathology, University of Rome " La Sapienza", Rome (Italy)

Summary

Teratomas are neoplasms composed of tissue foreign to the area in which it is found. They are considered to be an acquired neoplastic disease and familial incidence has not been reported. Only one occurrence of teratoma between monozygotic twins has been found in the literature. Here we report the case of two heterozygotic twins with benign cystic teratomas of the ovary as a base for future research for efficacy of an accurate familial follow-up in order to diagnose this neoplasm in early stage and for the molecular understanding of pathogenesis of teratoma.

Key words: Teratoma; Ovary; Twins; Familiarity.

Introduction

Benign cystic teratomas of the ovary are the most common of all ovarian neoplasms, representing approximately 20% of them. A review of the literature demonstrated only one previous report of a benign cystic teratoma of the ovary in two identical twins [1]. Here we report the case of two heterozigotic twins with benign teratomas of the ovary.

Case reports

Case 1.

L.L., a 30-year old woman, gravida 1 para 1 aborta 0, was admitted to the Department of Surgery for a right ovarian cyst on September 1994. The diagnosis was performed during a gynecologic check-up. Her past history was noncontributory. Her family history was remarkable: her mother had firstly undergone a right hemicolectomy for colon adenocarcinoma and secondly a partial cystectomy for bladder adenocarcinoma. Physical examination was negative. Pelvic ultrasonography revealed a right ovarian cyst (3.5 cm in diameter) with calcifications inside (Figure 1). The left ovary and the uterus appeared within normal limits. Radiographic diagnosis was a dermoid cyst of the right ovary. The left adnexa was normal. No lymphadenopathy or metastatic localization were found. Laboratory data on admission were within normal limits except for the presence of erythrocytes and leukocytes at urinalysis. On September 1994 she underwent surgical exploration. Laparoscopy revealed a right ovarian cyst and a salpingo-ovariectomy was performed. No peritoneal fluid was found. The excised ovarian mass was a unilocular cyst 2.8 x 3 x 3.5 cm in diameter weighing 80 g. Atypical mitotic figures were absent. Histologic diagnosis was benign teratoma of the ovary (Figure 2). The postoperative course was uneventful. The patient is alive and well seven years later.

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Case 2.

L.E., a 36-year old woman, twin sister of L.L., gravida 3 para 2 aborta 1, was admitted to the Department of Surgery for an enlarging abdominal mass on June 2001. Physical examination revealed a large mass filling the hypogastrium almost up to the umbelicus which was thought to be attached to the left ovary. Her past medical history was noncontributory except for tonsillectomy at the age of five. Ultrasonic tomography revealed a cystic neoplasm with septa and mild vascularization at power doppler. The neoplasm was 19 cm in diameter and appeared to be from the right ovary (Figure 3). The bladder and uterus were dislocated. Angio TC detected a voluminous mass (16 x 13 cm in diameter) in the right lower quadrant deforming the abdominal wall and the bladder (Figure 4). The neoplasm seemed fatfilled and well capsulated, not infiltrating neighbouring organs. Secondary mild hydroureteronephrosis on the right side was associated.

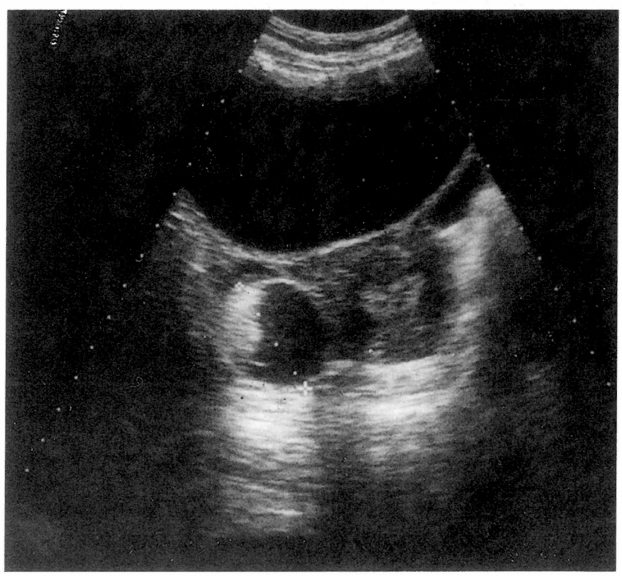

Figure 1. — *Case 1.* Pelvic ultrasonography showing a right ovarian cyst with calcifications.

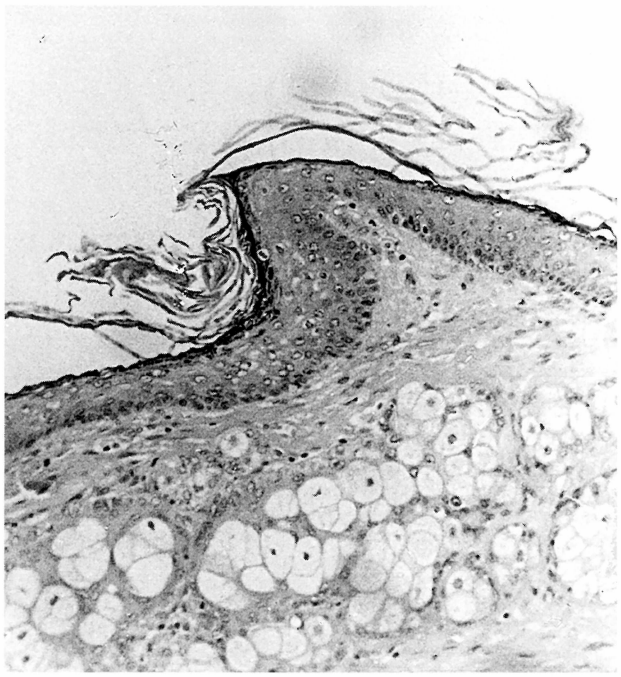

Figure 2. — *Case 1.* Histological features of the teratoma (magnification x100).

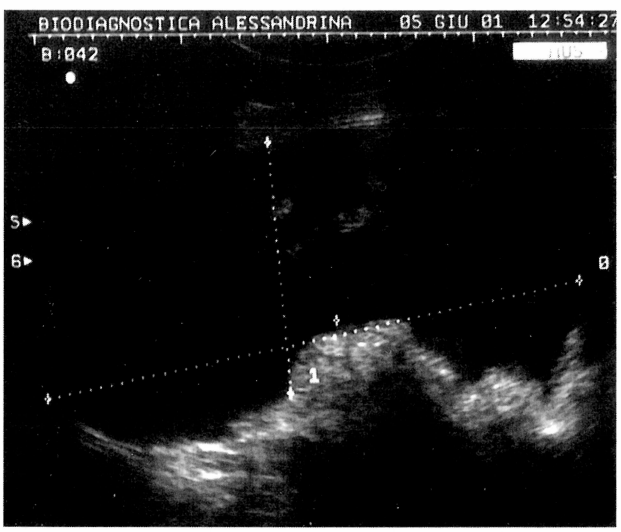

Figure 3. — *Case 2.* Ultrasonographic appearance of the ovarian teratoma.

Radiographic diagnosis was a dermoid cyst of the left ovary. The right adnexa was normal. No lymphadenopathy or metastatic localization were found. Laboratory data on admission were within normal limits except for CA125 dosage (CA125 43 U/ml, normal range < 35 U/ml). On June 2001 she underwent surgical exploration. Laparotomy revealed a large left ovarian mass that filled the abdomen, and 200 ml of transparent fluid were present in the peritoneal cavity. A left salpingo-oophorectomy was performed. The postoperative course was uneventful. Cytologic examination of the peritoneal fluid was negative for neoplastic cells. She is well 11 months after surgery. The excised ovarian mass was a unilocular fatfilled cyst 28 x 12 x 8 cm in diameter weighing 1,980 g. Atypical mitotic figures were absent. Histologic diagnosis was a benign teratoma of the ovary (Figure 5).

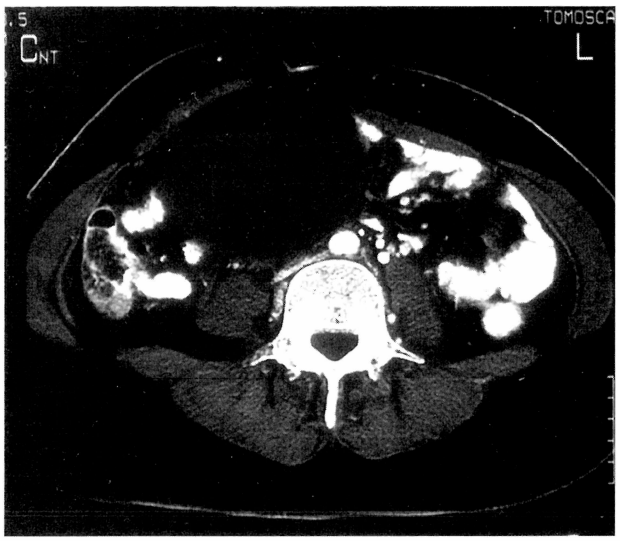

Figure 4. — *Case 2.* A voluminous mass in the right lower quadrant deforming the abdominal wall and the bladder is detectable at computed tomography.

Discussion

Germ cell tumors of the ovary are much less common than epithelial ovarian neoplasms and account for 2-3% of all ovarian cancers in Western countries. They occur more frequently in young women with a peak incidence in the early 20s, thus appropriate management by specialists is exceedingly important. There has been an important change over the past 20 years in the management of these neoplasms, with an increased tendency for ovarian preservation, as evidenced by more frequent use of cystectomy and a decrease in controlateral ovarian biopsy. Many

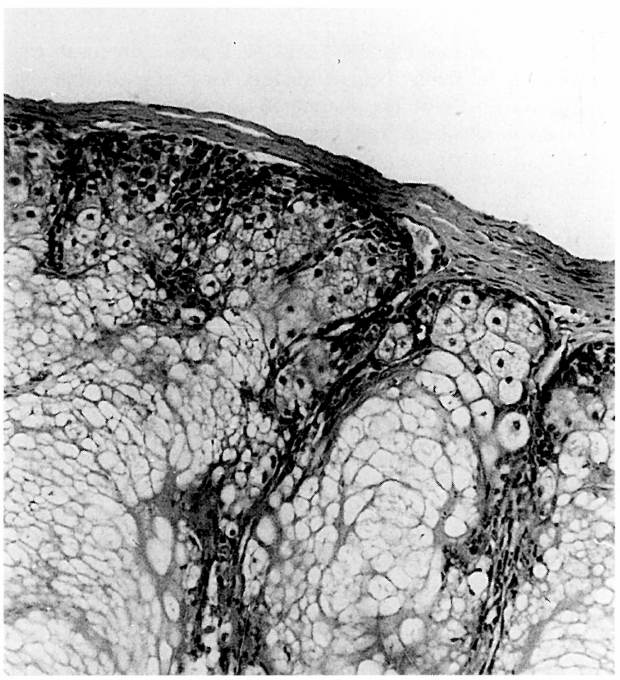

Figure 5. — *Case 2.* Histological features of the teratoma (magnification x100).

patients can have fertility preserved and the type of the surgical procedure will be dictated by operative findings [2-4]. Bilateral oophorectomy is not routinely necessary and the contralateral ovary and the uterus can be preserved. Germ cells tumors can be divided into dysgerminomas and nondysgerminomatous germ cell tumors. Among the latter teratomas are considered. A teratoma is a neoplasm composed of tissue foreign to the area in which it is found. Teratomas may be solid or cystic in appearance and are often referred to as dermoid cysts if unilocular. They contain elements from all three germ cell layers with a predominance of the ectodermal component in most tumors. The cause has not been identified. Environmental factors other than exposure to radiotherapy and radiation have not been documented. Two hypotheses can be contemplated. The first hypothesis supposes that totipotent cells are segregated in the development of the morula and then originate derivates of all germinal cell layers. Derivates are similar to that of the corresponding orthotopic tissues, even if in some cases morphology is much less well defined, attesting to the inductive effect of the contiguous tissues that are normally present in a specific organ [5-12]. The second hypothesis surmises that an unknown spark stimulates an asexual development or parthenogenesis. Mature cystic teratoma of the ovary is one of the most common ovarian tumors found in women of reproductive age. It represents 10-20% of all ovarian tumors in women during the second and third decades of life. Malignant transformation occurs at an incidence of approximately 2% [13-16]. Squamous cell carcinoma arising from the ectodermal layer is by far the most common cancer. Preoperative diagnosis of squamous cell carcinoma arising in the teratoma tissue is difficult and malignant degeneration is usually diagnosed postoperatively. Teratoma of the ovary is considered to be an acquired neoplastic disease and familial incidence has not been reported. Cases of familial gonadal cancers are reported, but in twins they are less frequent [1, 17-20]. Two extremely rare occurrences of testicular tumors in twins have been reported [21, 22]. Cobb described monozygotic twins with three different germ cell tumors [23]. Only one occurrence of teratoma between monozygotic twin sisters has been reported [1]. A review of the literature failed to demonstrate the incidence of benign cystic teratomas in twins to be different than that in offspring of single births and patients with teratoma have healthy children. This is the first report of two heterozygotic twins with metachronous teratoma of the ovary. We report the cases of our patients as a basis for future research in the molecular understanding and pathogenesis of teratoma, bearing in mind a possible familial incidence or an unknown environmental factor.

Although germ-cell tumors constitute less than 5% of all ovarian malignancies, they are important because they occur in young women, display a vastly different natural history than epithelial tumors, and require different treatment. Early diagnosis and treatment of teratoma can prevent the risk of malignant transformation, thus surgical intervention can be limited and most patients can retain their normal uninvolved ovary with preservation of normal menstrual and reproductive function.

References

[1] Brown E. H.: "Identical twins with twisted benign cystic teratoma of the ovary". *Am. J. Obstet. Gynecol.*, 1979, *134*, 879.

[2] Uznlar A. K., Yalinkaya A., Yaldiz M., Kilinc N., Gul T.: "Survival and reproductive function after treatment of immature ovarian teratoma". *Eur. J. Gynaecol. Oncol.*, 2001, *22*, 384.

[3] Elsheikh A., Milingos S., Kallipolitis G., Loutradis D., Vlachos G., Liapi A *et al.*: "Ovarian tumors in young females. A laparoscopic approach". *Eur. J. Gynaecol. Oncol.*, 2001, *22*, 243.

[4] El-Lamie I. K., Shehata N. A., Abou-Loz S. K., El-Lamie K. I.: "Conservative surgical management of malignant ovarian germ cell tumors: the experience of the Gynecologic Oncology Unit at Ais Shams University". *Eur. J. Gynaecol. Oncol.*, 2000, *21*, 605.

[5] den Boon J., van Dijk C. M., Helfferich M., Peterse H. L.: "Glioblastoma multiform in a dermoid cyst of the ovary. A case report". *Eur. J. Gynaecol. Oncol.*, 1999, *20*, 187.

[6] Ferrer J., Herrero-Zapatero A., Orille V., Llaneza P., Perez-Rodrigo C.: "Thyroid carcinoma originating in a teratoma of the ovary". *Eur. J. Gynaecol. Oncol.*, 1995, *16*, 396.

[7] Elemenoglou A., Zizi-Serbetzoglou A., Trihia H., Vasilakaki T., Bournia E.: "Mixed ovarian neoplasm composed of struma ovarii and Brenner tumour. Report of a case". *Eur. J. Gynaecol. Oncol.*, 1994, *15*, 138.

[8] Esik O., Nemeth G., Szepeshazi K.: "Calcitonin-secreting ovarian strumal carcinoid". *Eur. J. Gynaecol. Oncol.*, 1994, *15*, 211.

[9] Bjersing L., Cajander S., Rogo K., Ottosson U. B., Stendahl U.: "Glioblastoma multiform in a dermoid cyst of the ovary". *Eur. J. Gynaecol. Oncol.*, 1989, *10*, 389.

[10] Ajithkumar T. V., Abraham E. K., Nair M. K.: "Osteosarcoma arising in a mature cystic teratoma of the ovary". *J. Exp. Clin. Cancer Res.*, 1999, *18*, 89.

[11] Vadmal M., Hajdu S. I.: "Prostatic tissue in benign cystic ovarian teratomas". *Hum. Pathol.*, 1996, *27*, 428.

[12] Kuroda N., Hirano K, Inui Y., Yamasaki Y., Toi M., Nakayama H. *et al.*: "Compound melanocytic nevus arising in a mature cystic teratoma of the ovary". *Pathol. Int.*, 2001, *51*, 902.

[13] Wang P. H., Yen M. S., Juang C. M., Chen Y. J., Chao H. T., Yuan C. C.: "Intraperitoneal cancer spread after laparoscopic cystectomy for mature teratoma with malignant transformation". *Eur. J. Gynaecol. Oncol.*, 2002, *23*, 131.

[14] Kontogianni E., Koukoura E., Christopoulou E.: "Squamous cell carcinoma arising in mature cystic teratoma of the ovary: a case report". *Eur. J. Gynaecol. Oncol.*, 2000, *21*, 605.

[15] Comerci J. T. Jr., Jones J. G., Fields A. L., Runowicz C. D., Goldberg G. L.: "Squamous cell carcinoma in mature cystic teratoma in a young woman: a diagnostic and management dilemma". *Eur. J. Gynaecol. Oncol.*, 1996, *17*, 501.

[16] Moggian G., Saragoni A., Pellegri E., Tamburini E., Pini P. L., Giorgini G., Tumedei U. *et al.*: "Struma ovarii: report of a case with malignant evolution and of another observed in the course of pregnancy". *Eur. J. Gynaecol. Oncol.*, 1980, *1*, 201.

[17] Liber A. F.: "Ovarian cancer in mother and five daughters". *Arch. Pathol.*, 1950, *42*, 280.

[18] Lewis A. C. W., Clare Davison B. C.: "Familial ovarian cancer". *Lancet*, 1969, *2*, 235.

[19] Trentini G. P., Palmieri B.: "An unusual case of gonadic germinal tumors in a brother and sister". *Cancer*, 1974, *33*, 250.

[20] Musella R., Giaccone G., D'Ambrosio E., Calciati A.: "Father-son testicular cancer, case report". *Tumori*, 1983, *69*, 269.

[21] Levey S., Grabstald H.: "Synchronous testicular tumors in identical twins". *Urology*, 1975, *6*, 754.

[22] Ritter S.: "Twins with testicular tumors". *Med. Klin.*, 1992, *87*, 489.

[23] Cobb R. A., Fawcett D. P.: "Monozygotic twins with three different germ cells tumors". *Br. J. Urol.*, 1988, *61*, 262.

Address reprint requests to:
M. INDINNIMEO, M.D.
Via Cassia, 52
00191 Rome (Italy)