

Rare case of an ovarian monodermal teratoma with functional stroma and extensive ovarian decidualization in a 74-year-old woman

E. Vouza¹, Ch. Dastamani¹, Ch. Iavazzo², K. Bakalianou², D. Hasiakos², A. Kondi-Pafiti¹

¹Pathology Laboratory and ²2nd Department of Obstetrics and Gynecology, University of Athens, Aretaieion Hospital, Athens (Greece)

Summary

We present the clinicopathological findings of a rare case of a monodermal teratoma of the right ovary with functional ovarian stroma and extensive decidualization in a 74-year-old woman. The patient presented with vaginal bleeding. Ultrasound scan revealed a pelvic mass measuring 9.5 cm in the lower right abdomen. A right oophorectomy was performed. The tumor was cystic and multilocular filled with colloid material. Histological examination revealed follicles of thyroid type, and stromal clusters of fusiform or polygonal cells were found in the stroma. An extensive decidual reaction was observed. Morphological and immunohistochemical examination of the tumor revealed cystic struma ovarii with functional ovarian stroma and ectopic decidua. Total abdominal hysterectomy with oophorectomy was performed. A benign endometrial polyp, proliferative endometrium, two fibroids, and an ovarian cyst were observed.

Key words: Struma ovarii; Ovarian stroma; Decidua; Ovary.

Introduction

Mature teratomas represent 27-44% of all the ovarian tumors and are usually present during reproductive age, with a mean age at diagnosis of 32 years [1, 2]. Generally they are unilateral tumors and in 8-15% of the cases they are bilateral [1, 2]. Monodermal teratomas are rare tumors with the thyroid element as the main characteristic [1, 2]. Struma ovarii is found in 2.7% of all teratomas [1, 2]. It usually develops at the fifth decade of life [1], however some cases of struma ovarii in adolescents or in postmenopausal women have been reported in the literature [1, 3]. The clinical presentation is as an ovarian solid and cystic tumor and it rarely produces signs of hyperthyroidism. One third of the patients present with ascites or even Meigs syndrome [4, 5]. Decidualization is a normal finding during pregnancy, however it is not a common finding in postmenopausal women.

A rare case of an ovarian monodermal teratoma (struma ovarii) with functional ovarian stroma and extensive decidualization in a 74-year-old woman is presented.

Case Report

This is a case of a 74-year-old patient who presented to our department with vaginal bleeding of a month's duration. Blood tests revealed normal levels of estrogen, progesterone, free T3 and freeT4. No remarkable personal or family history was reported and the patient had not received any hormonal therapy in the past.

Ultrasound scan revealed a tumor measuring 9.5 cm in diameter in the left ovary and an endometrial polyp measuring 0.7 cm. The patient underwent exploratory laparotomy and after frozen section biopsy of the left ovarian mass showing thyroid tissue, an abdominal hysterectomy with the right adnexa was performed.

The gross examination of the ovarian mass showed a partly solid, partly cystic, multilocular tumor measuring 9.5 cm in the greatest diameter filled with colloid-like material. The cystic wall measured 0.1-1 cm. After routine procession of the specimen, formalin-fixed paraffin sections were stained with hematoxylin-eosin, and additional sections from the ovarian tumor and ovarian stroma underwent immunohistochemical investigation by a streptavidin-biotin method (Ventana, Benchmark). Histological diagnosis was **struma ovarii** (Figure 1). The residual ovarian stroma revealed cells with acidophilic cytoplasm like luteal or Leydig cells. Locally nodules of fusiform cells were found, resembling theca cells (Figure 2) with hyperplastic changes in loose edematous stroma. In the ovarian cortex, clusters of large cells with eosinophilic cytoplasm and a distinctive nucleus were found (Figure 3) resembling decidua cells.

Immunohistochemical analysis of the tumor cells showed a positive immunoreaction to thyroglobulin and TTF-1 and a negative immunoreaction to AE1/AE3 excluding adenocarcinoma. The decidua-like cells showed a positive immunoreaction to vimentin (Figure 4) and negative immunoreaction to CD-68 excluding the cases of histiocytic infiltration. The immunophenotype was consistent with the decidual cells, inspite of the paradox of the patient's age and absence of history of hormonal therapy of any kind. Furthermore, the stromal cells were positive for hormone receptors (ER, PgR) hCG and inhibin.

Histological examination of the uterus revealed an endometrial adenomatous polyp measuring 0.7 cm and two fibroids 0.5 and 2.5 cm in diameter. The endometrium was proliferative with mitotic activity and tubal metaplasia, despite the patient's age. An ovarian cyst was observed in the right ovary. No other therapy was considered necessary and the patient is well and without any tumor recurrence 48 months after surgery.

Revised manuscript accepted for publication August 5, 2010

Fig. 1

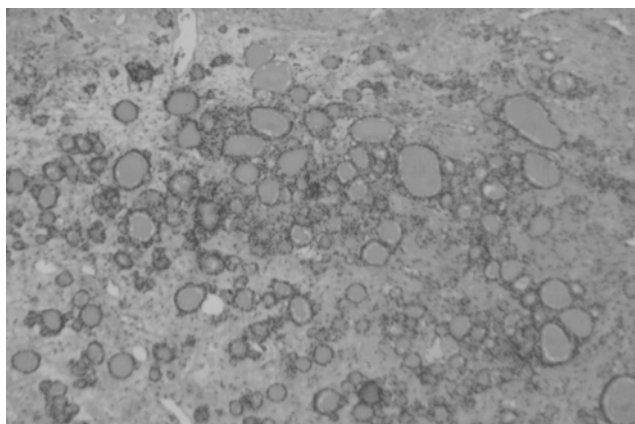


Fig. 2

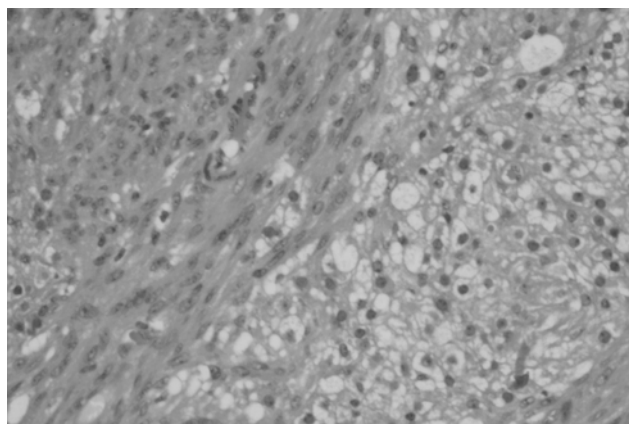


Fig. 3

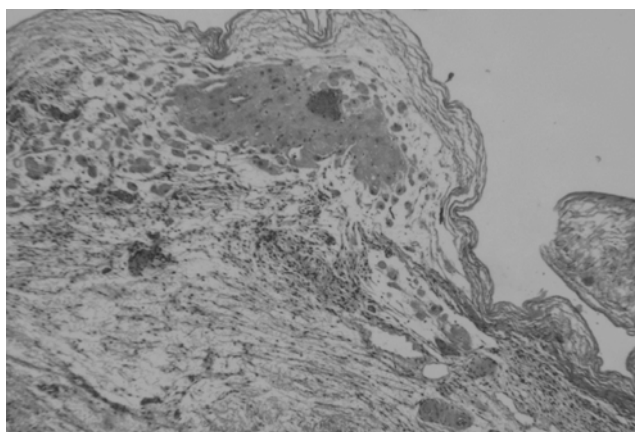


Figure 1. — Histological section of ovarian teratoma showing struma ovarii morphology, with small follicles filled with colloid (H&E x 25).

Figure 2. — Histological section of ovarian stroma showing hyperplastic theca cells in a loose stroma (H&E x 120).

Figure 3. — Histological section of ovarian cortex showing clusters of decidua-like large cells (H&E x 25).

Discussion

In our case the ovarian neoplasm presented the features of a monodermal teratoma of struma ovarii type. Most interesting are the histopathologic findings of hormone-producing stromal cells with extensive decidualization of the ovary.

Monodermal teratoma, struma ovarii, presents in the fifth decade of life as an abdominal mass in one-third of the cases with ascites, and rarely as Meigs syndrome, or with signs of thyrotoxicosis [1, 3-5]. Usually, it is a unilateral brownish tumor measuring 0.5-10 cm [1, 2]. Cystic struma ovarii is a multilocular cystic tumor measuring up to 20 cm, filled with colloid-like liquid that may lead to the diagnosis [1, 2]. Microscopic examination of cystic struma ovarii shows a thin fibroid wall with thyroid type follicles. Immunohistochemistry is positive for thyroglobulin and TTF-1 [6]. Those immunohistochemical findings aid in the differential diagnosis from Sertoli-Leydig tumors [6]. Histologic examination of struma ovarii may reveal various types of thyroid pathology, from normal or hyperplastic thyroid cells to adenoma or even carcinoma, usually of papillary type with the characteristic ground glass nuclei [7-9].

The differential diagnosis of functional ovarian stromal cells includes ovarian stromal reaction to mucinous neoplasms, Brenner tumors, other monodermal teratomas, dermoid cysts, dysgerminomas and less commonly

epithelial ovarian carcinomas or metastatic ovarian tumors [7-11].

Pregnancy is the main cause of ovarian decidualization. Less common causes are progesterone treatment, trophoblastic disease and hormone producing tumors of the ovaries or the adrenal glands. Rare causes are pelvic irradiation or as an idiopathic finding in pre- or postmenopausal women. However, it should be mentioned that ectopic decidua is a random histologic finding [12].

Conclusion

In our case a rare combination of monodermal teratoma of cystic struma ovarii and of functional ovarian stroma with decidualization was observed. The could be explained either as idiopathic or due to progesterone-producing stromal cells, as the patient did not report any other hormone-producing tumor or hormone treatment. The treatment of choice is total abdominal hysterectomy with bilateral oophorectomy.

References

- [1] Szyfelbein W.M., Young R.H., Scully R.E.: "Cystic struma ovarii: A frequently unrecognized tumor. A report of 20 cases". *Am. J. Surg. Pathol.*, 1994, 18, 785.
- [2] Iavazzo C., Vorgias G., Psarrou A., Lekka I., Katsoulis M.: "Late struma ovarii diagnosis many years after total thyroidectomy. A rare entity". *J. Buon.*, 2008, 13, 573.

- [3] Rutgers J.L., Scully R.E.: "Functioning ovarian tumors with peripheral steroid cell proliferation: A report of 24 cases". *Int. J. Gynecol. Pathol.*, 1986, 5, 319.
- [4] Guida M., Mandato V.D., Di Spiezio Sardo A., Di Carlo C., Giordano E., Nappi C.: "Coexistence of Graves' disease and benign struma ovarii in a patient with marked ascites and elevated CA-125 levels". *J. Endocrinol. Invest.*, 2005, 28, 827.
- [5] Bokhari A., Rosenfeld G.S., Cracchiolo B., Heller D.S.: "Cystic struma ovarii presenting with ascites and an elevated CA-125 level. A case report". *J. Reprod. Med.*, 2003, 48, 52.
- [6] McCluggage W.G., Young R.H.: "Immunohistochemistry as a diagnostic aid in the evaluation of ovarian tumors". *Semin. Diagn. Pathol.*, 2005, 22, 3.
- [7] Clement P.B., Young R.H.: "Tumor-like lesions of the ovary". In: Clement P.B., Young R.H (eds.). *Atlas of Gynecologic Oncology*, 2nd edition, Philadelphia, W.B. Saunders, 2008, 294.
- [8] Zhang X., Axiotis C.: "Thyroid-type carcinoma of struma ovarii". *Arch. Pathol. Lab. Med.*, 2010, 134, 786.
- [9] Szyfelbein W.M., Young R.H., Scully R.E.: "Struma ovarii simulating ovarian tumors of other types. A report of 30 cases". *Am. J. Surg. Pathol.*, 1995, 19, 21.
- [10] Tokunada H., Akahira J., Suzuki T.: "Ovarian epithelial carcinoma with estrogen-producing stroma". *Pathol. Internat.*, 2007, 57, 285.
- [11] Hayasaka T., Nakahara K., Kojimahara T.: "Endometrioid adenocarcinoma with a functioning stroma". *Obstet. Gynecol. Res.*, 2007, 33, 381.
- [12] Mils S., Carter D., Greenson J., Reuter V.E., Stoler M.H.: "Sternberg's Diagnostic Surgical Pathology". 5th edition, Lippincott, Williams & Wilkens, Philadelphia, 2010, 2333.

Address reprint requests to:
A. KONDI-PAFITI, M.D.
Pathology Laboratory
Aretaieion University Hospital
Vas. Sofias Ave, Athens 11528 (Greece)
e-mail: akondi@med.uoa.gr