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Rare Tumors, Rare Association: Ovarian Strumal Carcinoid - Retroperitoneal Cystic Lymphangioma

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Rezumat

Tumori rare, asociere rară: carcinoid ovarian strumal - limfangiom chistic retroperitoneal

Introducere: Carcinoidul strumal este un neoplasm rar de ovar care are o componentă tiroidiană (Struma) și o componentă carcinoidă confirmate prin imunohistochimie.
Prezentare de caz: O femeie de 55 de ani cu histerectomie și ovarectomie dreaptă pentru leiomiom uterin și chist ovarian drept, în antecedente, în urmă cu 12 ani, a fost internată în Clinica I Chirurgie, Spitalul Universitar Sf Spiridon Iași cu dureri în hipogastru, prezente de 2 luni. Testele de laborator au fost normale ca și testele serologice pentru markeri tumorali. Ecografia și CT au găsit pe topografia ovarului stâng o tumoră chistică, bine delimitată, cu o grosime neuniformă a peretelui, de 63/57/71 mm. Laparoscopia exploratorie, după adezioliză, a identificat o tumoră chistică retroperitoneală dezvoltată în mezosigmoid care a fost excizată, trompă uterină cu dilatare chistică și ovar stâng cu capsula intactă, pentru care am realizat anexectomie stângă cu extragerea pieselor tumorale în endobag. Evoluția postoperatorie a fost simplă, pacienta fiind externată după 3 zile postoperator. Examenele histopatologic și imunohistochimic au confirmat diagnosticul de carcinoid strumal ovarian și limfangiom chistic mezosigmoidian. Controlată la un an postoperator pacienta nu prezintă recidivă.
Concluzii: Această asociere între carcinoidul strumal ovarian cu limfangiomul chistic mezosigmoidian este neobișnuită și rară, nefiind citat în literatura de specialitate. Abordul laparoscopic pentru cele două tumori este fezabil, sigur și cu rezultate post-operatorii bune, imediat și la distanță.

Cuvinte cheie: carcinoid ovarian strumal, limfangiom chistic, abord laparoscopic

Abstract

Introduction: Strumal carcinoid is a rare neoplasm of the ovary containing thyroid (struma) and carcinoid component revealed by immunohistochemistry.
Case present: A 55-years-old woman with hysterectomy and right oophorectomy for uterine leiomyoma and right ovarian cyst, performed 12 years ago, was referred to the First Surgical Clinic, St. Spiridon University Hospital Iași with pain in hypogastrium present for 2 months. Laboratory tests are normal and serum testing for tumor markers is unchanged. Ultrasound and CT finds for topography of the left ovary a well-defined 63/57/71 mm cystic mass, with a non-uniform wall thickness. Exploration laparoscopy, after adhesiolysis, identifies a cystic tumor developed in mesosigmoid which was excised and a uterine tube with cystic dilatation and left ovary with intact capsule, for which we performed left adnexectomy and extracted pieces of tumors in endobag. Evolution is simple, the patient being discharged after 3 days postoperatively. Histopathological exam and immunohistochemistry revealed ovarian strumal carcinoid and cystic lymphangioma. After one year follow up the patient is disease free.
Conclusion: This association between an ovarian strumal

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carcinoid with mesosigmoidian cystic lymphangioma is unusual and rare, not cited in the literature. A laparoscopic approach for the two conditions is safe with good immediate and distant postoperative course.

Key words: ovarian strumal carcinoid, cystic lymphangioma, laparoscopy approach

Introduction

Strumal carcinoid is a rare neoplasm of the ovary, consisting of an intimate mixture of both elements. Struma and carcinoid being revealed by immunohistochemistry (ICH) (1). Primary carcinoid tumors of the ovary represent 5% of ovarian mature teratoma (2) and they derive from pluripotent germ cells (3). Most tumors are incidental findings by surgical exploration or by imagistics and sometimes they present symptoms of enlarging mass. In order to diagnose this rare tumor it is necessary a multidisciplinary team, surgeon, radiologist and pathologist (3). Our experience with symptomatic ovarian strumal carcinoid on the left ovary remaining after histerectomy (for uterine leiomyoma) associated with retroperitoneal cystic lymphangioma developed in mesosigmoid is worth sharing and explains the publication of this case.

Case report

A 55-years-old surgical postmenopausal caucasian woman with diabetes mellitus type II, arterial hypertension, and previous history with hysterectomy and right oophorectomy for uterine leiomyoma and right ovarian cyst, performed 12 years ago, was referred to the First Surgical Clinic, St. Spiridon University Hospital Iași with persistent pain of about two months in lower abdomen. Clinically a scar is present on the abdomen after Pfannenstiel incision, fat is well represented, deep pain on palpation in hypogastrium and left iliac fossa. Laboratory tests are normal and serum testing for tumor markers is unchanged.

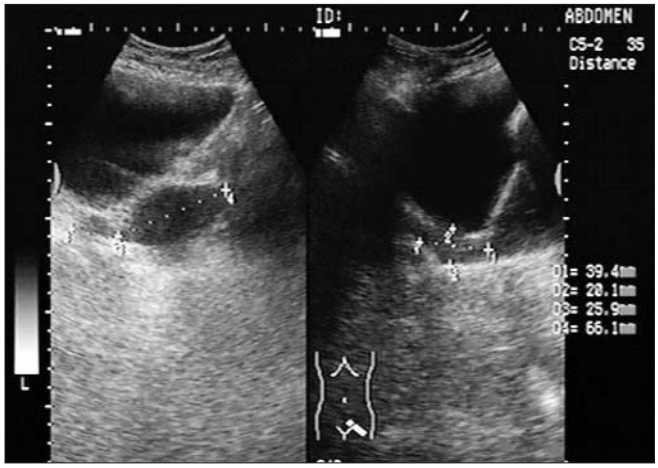

Figure 1. *Ultrasound findings: In hypogastrium a 58/22/59 mm hypoechogenic mass and in left side a 59/39/41 mm hypoechogenic mass, possible ovarian cyst*

Ultrasound revealed in hypogastrium a 58/22/59 mm hypoechogenic mass and in the left side area a 59/39/41 mm hypoechogenic mass, possibly an ovarian cyst (Fig. 1). Iodine contrast enhanced CT (Computed Tomography) finds for topography of the left ovary a well-defined 63/57/71 mm cystic mass, with a non-uniform wall thickness, posterolaterally reaching 14 mmm, with easy plug contrast and incomplete septum inside. The mass comes in contact with the superior ovarian vessels. Lower, the mass is adjacent to the vagina, anterolaterally right on a small area of sigmoid or ileal loops and on the left side widely adjacent to the parietal pelvic peritoneum (Fig. 2).

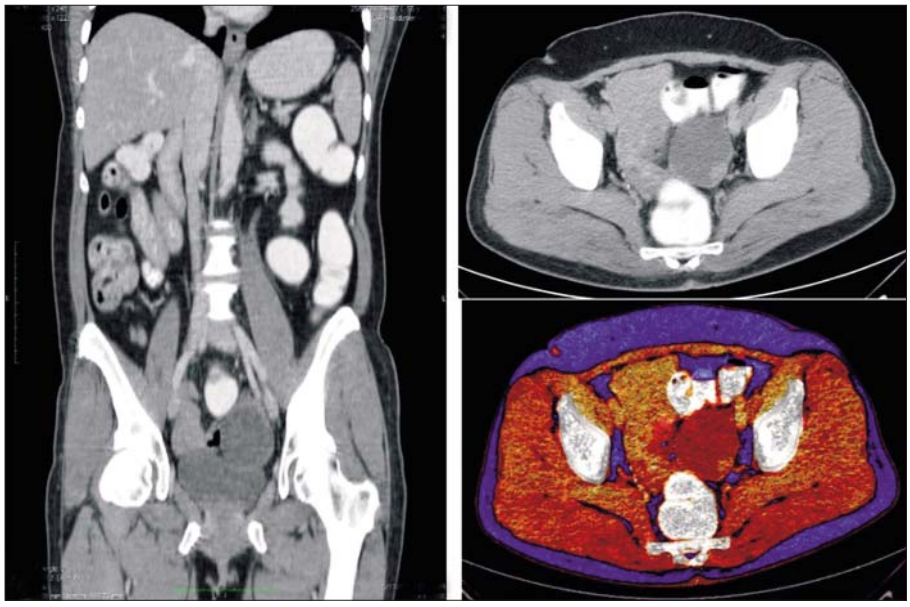

Figure 2. *CT findings - for topography of the left ovary a 63/57/71 mm well-defined cystic mass, with a non-uniform wall thickness in contact with the superior ovarian vessels, lower is adjacent to the vagina, anterolaterally right on a small area of sigmoid or ileal loops and on the left side widely adjacent to the parietal pelvic peritoneum*

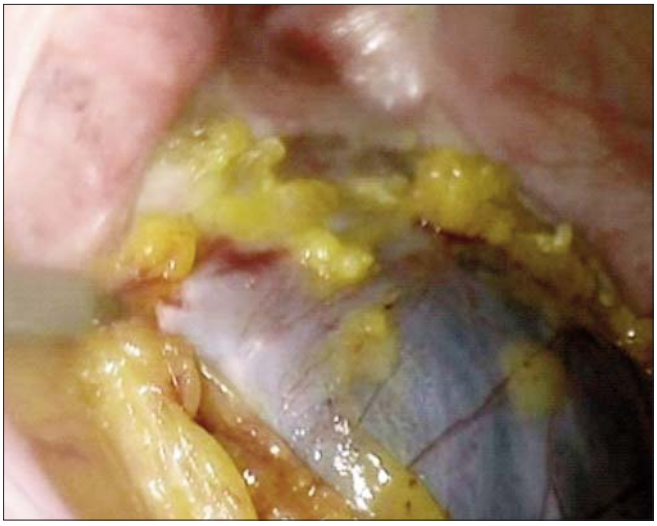

Figure 3. *Intraoperatory view - cystic lymphangioma developed in mesosigmoid*

We performed a laparoscopy exploration of the abdominal cavity identifying postoperative epiplono parietal adhesions and a cystic tumor developed in mesosigmoid which are excised (Fig. 3) and a uterine tube with cystic dilatation and left ovary with intact capsule and for which we performed left adnexectomy and extracted pieces of tumors in endobag (Fig. 4).

Evolution is simple, the patient being discharged after 3 days postoperatively.

Histopathology examination revealed, on gross examination after the cut sections, a yellowish brown tumor, arising from the left ovary, of firm consistency and intact capsule, with well-defined nodularity, of 1 cm and typical smooth cyst wall for lymphangioma.

Microscopy in left ovary showed abnormal thyroid follicles with colloid-like material (Fig. 5), mixed with a carcinoid component consisting of endocrine cells with trabecular disposal located in a fibrous stroma, suggestive for ovarian strumal carcinoid. A mesosigmoidian cyst is formed from vascular structures with cystic ectasia, some with open lumen, others containing hematic and isolated lymphoid infiltrates parietally, corresponding to a cystic lymphangioma issues (Fig. 6).

Immunohistochemical staining was positive for neuron-specific enolase, synaptophysin, and chromogranin, negative for calcitonin and Ki 67 < 1% (Fig. 7). The imunhistochimical profile corresponds to a benign tumor with differentiated thyroid and neuroendocrine carcinoid type.

After one year follow up, the patient is disease free, thyroid scintigraphy is normal and abdominal ultrasound did not reveal changes. The patient is monitored clinically-biologically by an endocrinologist.

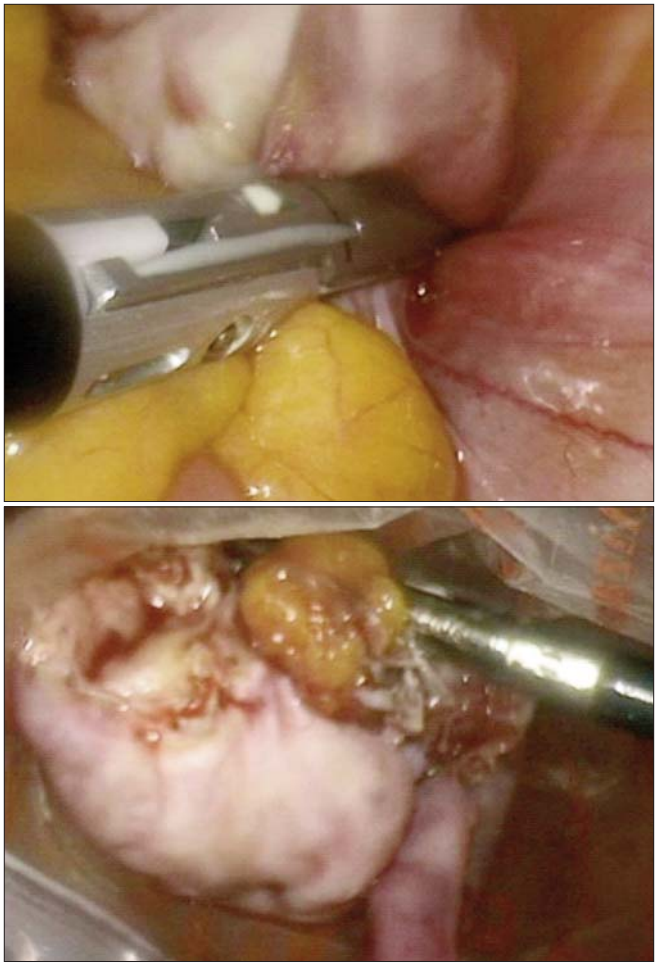

Figure 4. *Intraoperatory view - left adnexectomy with extracting pieces of tumors in endobag*

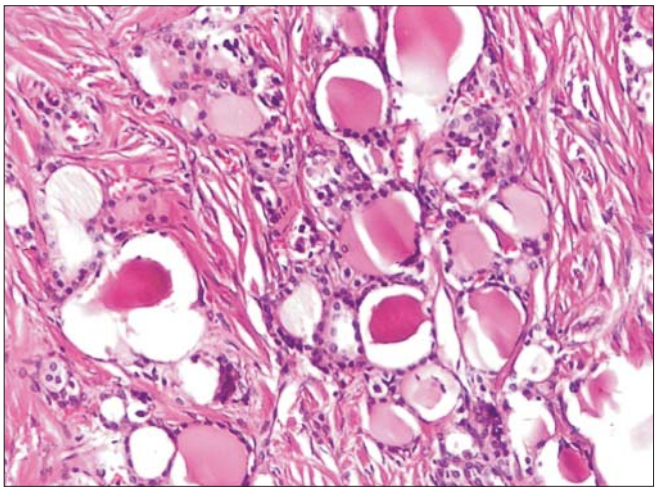

Figure 5. *Microscopy in left ovary showed abnormal thyroid follicles with colloid-like material (HE col., x 40)*

Discussions

Strumal carcinoid is an unsual neoplasm of the ovary containing different proportions of both components monodermal teratoma (struma ovari) and somatic tumors (primary ovarian carcinoid) (2,3).

Primary ovarian carcinoid tumors are extremely rare, accounting for 0.1% of all ovarian malignancies. As for as the

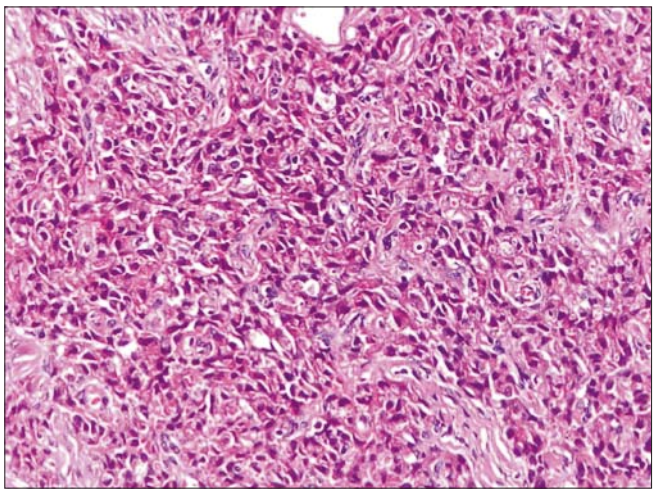

Figure 6. *Microscopy in left ovary showed a carcinoid component consisting of endocrine cells with trabecular disposal located in a fibrous stroma (HE col., x 10)*

number of carcinoids is concerned, they have been identified in 0.52% cases in USA and in 1.7% in Japan (1).

Although classified by WHO as a form of carcinoid, it can be interpreted as an associated ovarian struma with a secondary somatic neoplasm. The component struma is required to diagnose an ovarian carcinoid strumal.

The pathological classification of ovarian struma may include:

- Typical ovarian struma
- Variants with malignancies: strumal carcinoid, Brenner tumors, mucinous cystadenoma;
- Variants with thyroid cancer: papillary, follicular, insular carcinoma (3).

Macroscopically and microscopically, the thyroid and the carcinoid component may be distinct or mixed. Even in cases where the two components are separate at their interface there is often a mixture of these elements, suggesting that the carcinoid tissue occurs from struma. The thyroid tissue may resemble normal thyroid, with a colloidal goiter, a microfolicular or macrofolicular adenoma or can take the appearance of papillary and follicular carcinoma.

The carcinoid neuroendocrine component looks trabecular (50% of cases), insular or present a mixed model (1); only a third of the insular tumors may present symptoms of carcinoid syndrome (1).

The thyroid component of strumal carcinoid can express thyroxine, TTF - 1 and rarely, calcitonin (4). The neuroendocrine component of carcinoid stromal is frequently positive for chromogranin, somatostatin and serotonin (3). Sometimes the trabecular carcinoid may accompany severe constipation, explained by the production of peptide YY (5, 6); constipation disappears after removal of the tumor.

The disease occurs most frequently in the fifth decade, although cases were reported where it which occurred at puberty and menopause. In most cases these tumors are found accidentally in peri-or postmenopausal woman. The clinical aspect is uncharacteristic: small pelvic tumor, rarely ascites, and carcinoid syndrome or thyroid disorders (2).

The diagnosis of strumal carcinoid involves a multidisciplinary collaboration (surgeon, radiologist and pathologist) and must be confirmed by IHC (7).

The differential diagnosis of strumal carcinoid includes the other forms of carcinoid tumours, especially the trabecular, a type of cancer that is similar to carcinoid strumal, but without containing thyroid follicles.

In older cases from literature, strumal carcinoids were

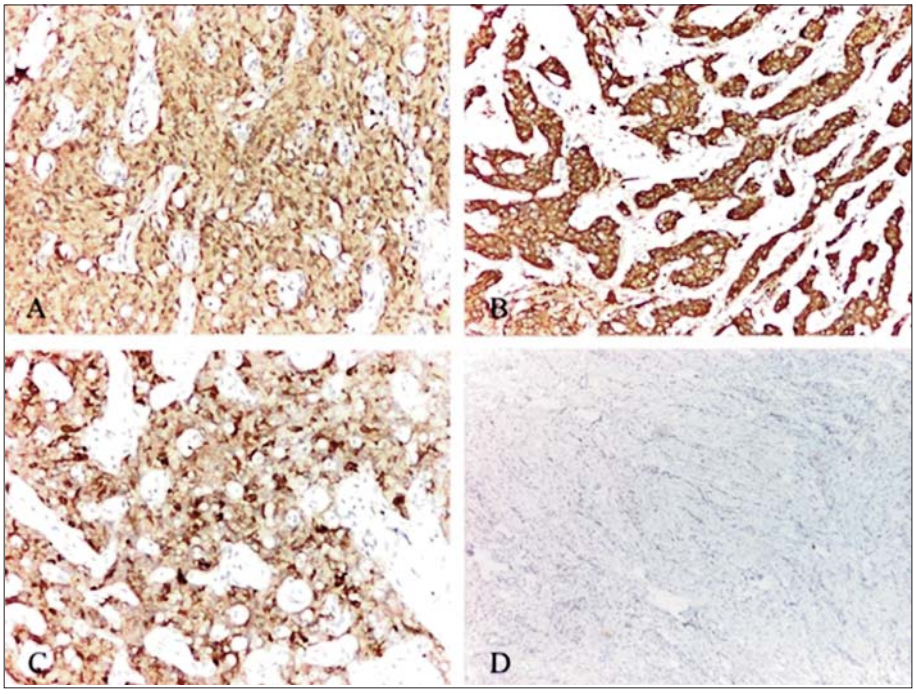

Figure 7. *Immunohistochemistry – positive for neuron-specific enolase (A – ob. x4), synaptophysin (B – ob. x4), and chromogranin (C – ob. x4), negative for calcitonin (D – ob. x4) corresponds to an ovarian strumal carcinoid*

misdiagnosed as malignant struma ovarii, although the histological appearance of strumal carcinoid isn't very similar to the various types of thyroid carcinoma.

Tamsen A and Mazur MT (8) reported the case of a patient with multiple endocrine neoplasia type 2A (syndrome Sipple). The case we report refers to a particular association of ovarian strumal carcinoid with cystic lymphangioma of mesosigmoid.

The treatment of these tumors is primarily surgical. A laparoscopic approach has more frequently been used in surgical practice, since 1997 with good long-term results (9-11).

For strumal carcinoid tumors in stage I, unilateral or bilateral oophorectomy with or without hysterectomy, are recommended. Davis KP et al. reported an excellent overall survival in the patients whose disease was confined to one ovary (100% 5-year survival), but only 1 of 6 patients survived (33%, 5-year survival) if in an advanced stage at diagnosis (12).

Postoperative course is generally favorable. Long-term prognosis is also favorable. From a series of 50 patients with ovarian strumal carcinoid tumor only a single relapse may be reported 18 months after surgery (13).

In another study by Talerman et al, a large number of 150 primary ovarian carcinoid tumors revealed that these have presented an ovarian low malignant potential (14), metastases are rare, localized in the lungs, breast (2% of cases) (15,16). In the presence of metastases confined to one of the components, especially the thyroid, thyroidectomy and treatment with I131 is indicated, which associated sometimes can lead to regression of metastatic disease (17). Unfavorable prognosis could be betrayed by IHC positive for topoisomerase II alpha and Ki-67. (18).

Abdominal cystic lymphangioma are mesodermal tumors resulting from a sequestration of lymphatic tissue derived from the retroperitoneal bags; as they are notable to establish communication with the venous system, they retain the power to bud proliferation and secretion of vascular endothelial turning into genuine tumors of lymphatic origin. Regardless of their congenital, retention or neoformation, the abdominal cystic lymphangioma betrays a causal relationship with the extensive retroperitoneal lymphatic network (19).

Most of them are asymptomatic, being discovered incidentally during imaging or surgery or complications (superinfection, compression, intracystic hemorrhage, rupture).

Ultrasound is specifically highlighting hypo- or anechoic masses encapsulated, uni- or multilocular, with thin-walled, transonic content, which may become hyperechoic in case of intracystic bleeding. Abdominal CT examination evaluates the size, the appearance of the lesions and the anatomical relationships lesion with adjacent organs. MRI is the gold standard that allows a better delimitation of the lesion and revealing complications, and especially the presence of intracystic bleeding (19). The laparoscopy is useful both in diagnosis of giant cystic masses with a preoperative undiagnosed origin and in treatment such as the one described (20).

The treatment is exclusively surgical. LC complete excision is almost always possible in uncomplicated cases. The recurrences occur after incomplete resection of the tumor. The laparoscopic approach should be the gold-standard. Depending on the location and size of the cystic lymphangioma total laparoscopic cystectomy without puncture (closed system) can be employed in the case of small cyst or after evacuation in the case of large cysts.

Conclusions

This association between an ovarian strumal carcinoid with mesosigmoidian cystic lymphangioma is unusual and rare, not cited in the literature. It may be a rare histogenetic anomaly encountered in the case of adults. A laparoscopic approach for the two conditions is beneficial, with a good postoperative course immediately and at distance.

Conflicts of interest

Authors have no conflicts of interest to disclose.

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