

Malignant Struma Ovarii: A Case Report**Zineb Chaqchaq***, MD Imane Benchiba, Ezza Lemrabott, Nisrine Mamouni, Sanaa Errarhay, Chahrazed Bouchikhi, Abdelaziz Banani

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Abstract: Struma ovarii is an ovarian teratoma which predominantly contains thyroid tissue. Because of its rarity, the diagnosis and the management of the tumour have not been clearly defined. We report a case of malignant papillary struma ovarii not suspected in preoperative. The surgical treatment of our patient was total hysterectomy, bilateral salpingo-oophorectomy and biopsies completed with thyroidectomy and radioactive iodine ablation which was decided in multidisciplinary consultation meeting. Through this observation, we discuss the diagnostic, management and outcome features of these particular tumors.

Keywords: Struma ovarii, malignant, surgery, radioiodine therapy.

INTRODUCTION

Teratomas are tumors of the germ line composed of cells derived from one or more of the three embryonic layers (meso, endo and ectoderm) that can be uni or multilocular. Ovarian goiters (GO) are ovarian uni-tissue teratoma composed predominantly (more than 50%) or exclusively (pure GO) of thyroid tissue. We report the case of papillary malignant ovarian goitre not suspected preoperatively; our objective through this observation is to discuss the anatomico-clinical and therapeutic particularities of this rare tumor.

CASE REPORT

A 50-year-old patient, single-aged, follow up for schizophrenia, presented a month before her admission a deep vein thrombosis of lower limb for which she was treated with anticoagulant.

She consulted in our institution for a significant increase in the abdominal volume evolving since 1 year. She also complained of abdominal pain associated with metrorrhagies and constipation. Clinical examination revealed the presence of a huge abdominal pelvic mass reaching the xiphoid appendix and having a renitent consistency.

Pelvic echography showed a large-scale abdominopelvic mass of massive size with full endocystic buds and non-dopplerized thickened septa.

A complementary pelvic thoraco-abdominal scanner has shown a large abdominal pelvic mass predominantly fluid ovarian, pushing the digestive tract towards the periphery without any repercussions on the urinary system.

Operative exploration showed a massive solidocystic adnexal right mass, making approximately 30cm. This mass presents a significant peripheral vascularization and does not contain exocystic vegetation. It was adherent to the fallopian tubes and enclaved at the level of the broad ligament. The rest of the exploration is unremarkable, including a normal-sized uterus with a macroscopically normal left appendage.

A right adnexectomy was performed and referred for extemporaneous examination which showed an evocative aspect of teratoma (ovarian goitre) or serous borderline epithelial tumor. A complementary total hysterectomy associated with left adnexectomy and multiple biopsies in the left and right paracolic gutters, and in the omentum was performed.

The definitive anatomopathological examination of the right adnexectomy shows macroscopically a solidocystic mass of 30 cm long axis. Microscopically, the ovarian parenchyma is the seat of a proliferation made of thyroid vesicles of

variable sizes, bordered by a regular foundation of thyreocytes. The tumor cells are increased in size, with overlapping nuclei marked by condensation of chromatin on the nuclear membrane producing a "frosted glass" appearance.

The diagnosis of carcinogenic papillary ovarian goitre has been retained. Peritoneal fluid sampling, total hysterectomy, left adnexectomy and various biopsies showed no histological abnormalities.

Biological exploration of the thyroid gland and a thyroglobulin testing were normal. A cervical

ultrasound showed a right thyroid nodule, which can be classified as 4A of the TIRADS classification.

This case was discussed in a multidisciplinary consultation meeting with the decision to complete the treatment with thyroidectomy associated with a radioiodine therapy.

The anatomopathological result of thyroidectomy showed thyroid hyperplasia with no malignancy.

Currently, the patient is in euthyroidie under levothyrox and has started the radioiodine therapy.



Fig-1: Ultrasound images showing a predominantly cystic abdominopelvic mass occupying the entire screen with endo-cystic buds and thick septa

DISCUSSION

Malignant ovarian goitre is a rare ovarian tumor. It represents 5% of all teratomas and occurs in the 5th decade [1]. Most often unilateral in more than 90% of cases (left ovary reached in 63% of cases) [2]. Metastatic spread is difficult to evaluate because of the rarity of the tumor; it is exceptional and estimated at less than 5% of cases [2].

Clinical symptomatology is usually poor. The diagnosis is made either in front of a systematic examination, or before the existence of an abdominal pelvic mass, of pelvic pain, ascites or sometimes hydrothorax [3, 5]. Signs of hyperthyroidism can be seen in 5 to 8% of cases [1] (ectopic thyroid tissue may become autonomic). The radiological appearance of ovarian goiters is not very specific and is often suspected of malignancy [2]. Ultrasound found a heterogeneous solido cystic tumor with septum and vegetation. The Doppler mode [4] shows a blood flow recorded at the center of the tumor lesion with a low resistance index due to the richly vascularized nature of the thyroid tissue).

In MRI it is a multi-lobulated complex mass with thickened septa, multiple cysts of variable signal

intensities with solid components [2]. The solid portions that enhance strongly after gadolinium injection correspond to thyroid tissue and stroma containing abundant blood vessels and fibrous tissue [1,6].

Biological assays [2] are not very specific for the diagnosis of ovarian goiters. In case of thyroid tumor hypersecretion, we find a peripheral hyperthyroidism profile with a decrease TSHus and increase T3 and T4. Ca125 is classically normal and does not constitute a marker of malignancy of ovarian goitre. We rarely find an increase in case of ascites or pleural effusion.

The diagnosis of ovarian goiters is histological. Macroscopically, the size of the tumor varies between 1.5 and 38 cm [7]. It is characterized by cavities with greenish or red-brown gelatin content sometimes in a dermis cyst [7, 1]. Its malignant transformation must be suspected in the presence of a solid zone often inconstant. The histological diagnosis of malignancy has long been controversial because of the lack of uniform diagnostic criteria and the rarity of the tumor.



Fig-2 and 3: Scanning sections showing voluminous abdominopelvic mass probably right ovarian predominantly fluid

Currently the consensus is to retain as criteria of GO malignancies the histological pathological features of primary thyroid cancer [1,8]. The cytological signs of malignancies are irregular cell nuclei, overlapping and frosted glass, intense mitotic activity, and vascular invasion signs. GOs are most often papillary carcinomas (70%) and more rarely follicular carcinomas [1]. IHC using TTF-1 (thyroid transcription factor1), thyroglobulin, and neuroendocrine markers can assist diagnosis [9, 1]. Due to the rarity of ovarian goiter, there is no consensus on management [2]. The treatment is based on limited cases reported in the literature [1].

Several authors advocate treatment of thyroid cancer type rather than ovarian cancer [3]. Ovarian surgery depends on the age of the patient and the locoregional extension [3]. We opt for a conservative attitude (unilateral adnexectomy) in a young patient desirous of pregnancy, if the tumor is localized and in the absence of capsular invasion [10]. A study by

Desimone *et al.* favors conservative treatment for patients wishing to become pregnant [11, 12]. At a later age and in case of tumoral extension, the treatment is radical. Radical treatment consists of total hysterectomy, bilateral adenotomy and complete peritoneal starvation including peritoneal fluid cytology and omentectomy [1, 13]. Para-aortic and pelvic lymphadenectomy is recommended for most authors because of the classic lymphophilic character character of thyroid cancers [3].

Supplementation by total thyroidectomy is systematically performed by some authors [11] and remains reserved for extended and metastatic forms for others. It is primarily indicated for the removal of ovarian metastatic primary thyroid tumors, for complementary radioactive iodine treatment, and for subsequent monitoring [3]. Desiman *et al* advocate total thyroidectomy for all patients. They justify their therapeutic attitude by the fact that in their review of the literature, all the patients who benefited from a

complementary treatment by radioactive iodine are in remission whereas 50% of the patients treated by ovarian surgery recidivated [11].

However, a thyroid tumor metastasized to the ovary, can be ruled out by a thyroid clinical examination and a thyroid ultrasound. A metastatic ovarian goitre must be bilateral and the histological study must not have shown other teratomatous components [14,1].

Adjuvant treatment with radioiodine therapy is indicated in case of locoregional tumor extension, distant metastasis or tumor recurrence [2]. Some authors recommend the administration of iodine 131 in a systematic way [11], others advocate, in the absence of a pejorative criterion of locoregional extension, a simple treatment as a treatment adjuvant [15].

Postoperative monitoring is identical to that of thyroid cancers [11]. This is done by clinical examination and by the measurement of thyroglobulin which is a marker of tumor recurrence or distant metastasis. In case of increase of the thyroglobulin the realization of a whole body iodine scintigraphy is indicated. Pelvic ultrasound can be used to check for locoregional recurrence [2].

The prognosis is difficult to assess because of the rarity of these tumors [1]. The localized forms have a very good prognosis [2]. Size, tumor adhesion or the presence of ascites would be less prognostic factors [15]. Makani *et al.*, in their review of the literature estimate the risk of recurrence at 15%, occurring on average 4 years after the initial diagnosis [13]. Very late metastases in summer reported by some authors. However, even metastatic forms are compatible with prolonged survival estimated at an average of 8.3 years [3].

CONCLUSION

Malignant ovarian goitre is a rare ovarian tumor whose clinical symptomatology is not very specific. The diagnosis is histological. Surgical treatment consists of unilateral adnexectomy, hysterectomy with bilateral adnexectomy, or hysterectomy bilateral adnexectomy omentectomy and lymphadenectomy. Treatment can be done by thyroidectomy combined with irathérapie.

Considering the rarity of the tumor and lack of consensus concerning the management, the therapeutic decision must be made in a pluridisciplinary consultation meeting.

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