Case Report

Struma Ovarii Mimicking Ovarian Carcinoma: a Case Report and Review of the Literature

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Abstract

Struma Ovarii is a rare neoplasia. It is a monodermic mixed teratoma, with predominance of thyroid tissue and represents 3% of ovarian teratomas. This article reports a case of Struma Ovarii in a 66 years-old patient, with a progressive abdominal mass, ascites and high levels of CA-125. The findings were highly suggestive of ovarian carcinoma. The CT scan showed a complex ovarian lesion and the patient was submitted to an exploratory laparotomy. The pathology report showed a left ovary Struma Ovarii.

Keywords: Struma Ovarii. Ovarian Neoplasms.

Introduction

Struma ovarii is a rare neoplasia. It consists of a thyroidal tissue monodermic teratoma, corresponding to about 3% of mature teratomas.[1] It generally affects patients with advanced age, being the peak of incidence the fifth decade of life (42 years is a mean age of incidence).[1,2] It rarely occurs in puberty. Most patients present an asymptomatic pelvic mass or abdominal pain, ascites and rarely hyperthyroidism.[1] This tumor is generally benign, although malignant transformation has been described in 5%-37% of cases,[1] corresponding to nearly 0.01% of malignant ovarian tumors,[1] and even when these tumors are malignant, metastases appear in only 5% to 6% of cases.[2] Most tumors are unilateral, and affect mostly the left ovary (nearly 63% of cases),[1-3] Bilateral lesions may take place in up to 6% of cases.[1]

Described for the first time by Von Kalden in 1895, Struma Ovarii is a unitissular teratoma of the ovary composed mostly (more than 50%), and sometimes exclusively (pure Struma Ovarii) by thyroidal tissue.[2]

Generally, a complex ovarian mass, associated to ascites, pleural effusion and CA-125 elevation, in post-menopausal patients leads the doctor to diagnose a malign neoplasia of the ovary. Ascites may be present in up to a third of cases.[4] But only a few cases in the literature report the association of ascites to CA-125 elevation.[4]

The present work reports a clinical condition of pelvic mass, ascites, pleural effusion and CA-125 elevation, frequent in ovarian cancer, in a patient with benign Struma Ovarii.

Case Report

A black Patient, 66 years, consulted the Service of General Surgery of Waldemar de Alcantara Hospital reporting an abdominal mass that had appearing one month before, with a progressive growth associated to increase of abdominal volume and dyspnea. She reported

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no hormonal complaints. An abdominal ultrasound showed ascites and a complex lesion in the left annex, having 5.0 x 3.5 x 1.0cm. Computed tomography of the abdomen and pelvis showed an important ascites, a small bilateral pleural effusion and a complex heterogeneous mass in the left ovary (Figure 1). The patient had hepatic and renal functions in normal levels and CA-125 higher than 600u/ml, besides 437 u/ml lactic desidrogenase.

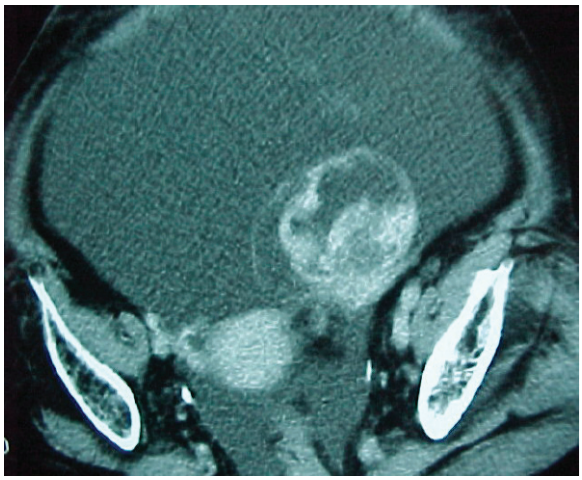

Figure 1 - Image of Computed Tomography of the Abdomen and Pelvis: important ascites, small bilateral pleural effusion and a complex heterogeneous mass in the left ovary

Before a condition suggestive of advanced ovarian cancer, an exploratory laparotomy with trans-operative anatomopathological evaluation was done. During the procedure, drainage of six liters of ascitic fluid was done and a nodular lesion in the left ovary, capsulated, with a solid and cystic component of about 5cm in the higher diameter, with a violaceous content inside was identified (Figures 2 and 3). The uterus, the right ovary, the epiplon, the parietal and visceral peritoneum, pelvic and paraaortic lymphonodes presented no macroscopic evidence of disease. The operative specimen was examined and showed a possible malignant complex lesion. The patient was submitted to hysterectomy with bilateral salpingooforectomy. Surgical procedure was performed without intercurrences and the patient was discharged in the second post-operative day, with resolution of ascites and normalization of CA-125.

Final result of the pathology showed a lesion of 5.0 x 4.0 x 3.8cm in the left ovary, solid, with a smooth and grayish external surface. When cut, it presented irregular cavities with a smooth internal surface of serous content interspersed by brownish tissue (Figure 3). After being paraffined, the specimen was examined and showed a lesion reproducing macro and micro thyroidal filaments containing colloid and birefringent concretions of calcium oxalate, a characteristic of Struma Ovarii (Figure 4). Immunohistochemistry showed positivity for cytokeratin 7, thyreoglobulin and TTF1, and negativity for CA-125.

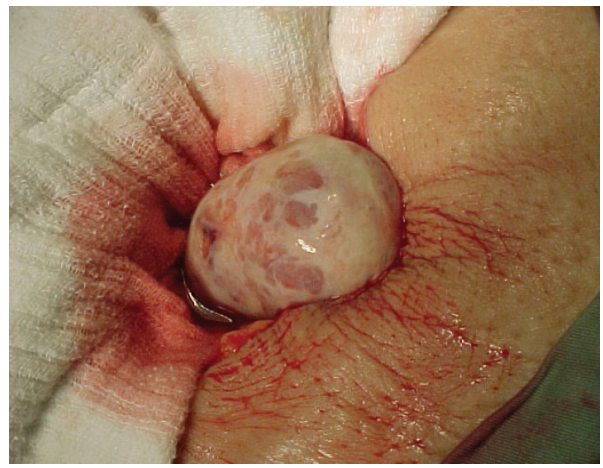

Figure 2 – A nodular lesion in the left ovary, capsulated, with a solid and cystic component of about 5cm in diameter, with a violaceous content inside

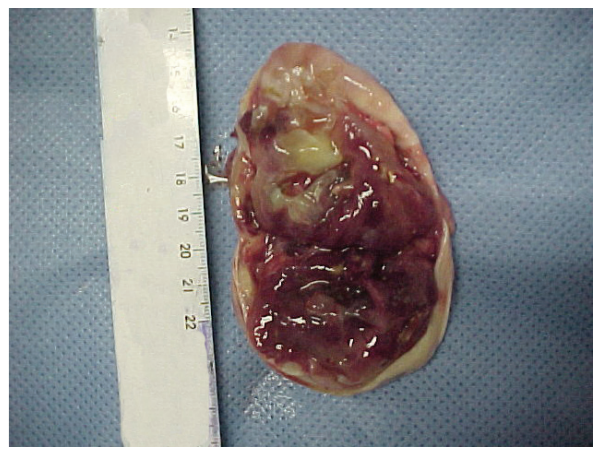

Figure 3 – An lesion of 5.0 x 4.0 x 3.8cm in left ovary, solid with smooth and grayish external surface. Cuts showed irregular cavities with smooth internal surface of serous content interspersed by brownish tissue

Discussion

Struma ovarii is an ovarian teratoma composed completely or predominantly by mature ovarian tissue and is generally unilateral, but may be bilateral in up to

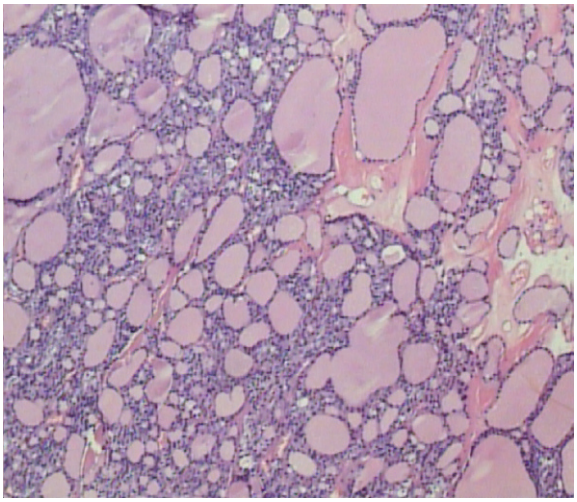

Figure 4 - Lesion reproducing macro and micro-micro thyroidal filaments containing colloid and birefringent concretions of calcium oxalate, a characteristic of Struma Ovarii

6%[1] of cases.

Teratomas are germinal-lineage tumors composed by derived cells of one or the three embryonic layers (ectoderm, endoderm mesoderm), and they may be unitissular or pluritissular.[2] Approximately 14% of teratomas have small focuses, very often insignificant, of thyroidal tissue. Around 5% to 8% can have clinic hyperthyroidism[2] and 25% to 33 % presents some degree of functional abnormality of the thyroid.[3]

In nearly 15 % to 20 % of the cases the Struma Ovarii it is associated to the ascites.There is no consensus on the etiology of the ascites in these cases. Among several hypotheses, it is described that the ascites can be secondary to the peritoneal irritation for the tumor or twist of the ovary, or for lymphatic obstruction, or for liberation of toxins and products. Other hypotheses suggest the formation of cysts inside the neoplasia like as an etiological factor for the ascites, while other authors suggest a discrepancy between the arterial and veined supply with impairment to the lymphatic drainage and, finally, ascites can be related to hypoalbuminemia. Without taking into account the etiology, ascites is very common in cancer, with positive predicted value of 95 %.5

Bulky ascites also was the finding in the case described by Huh et al.,[6] which suggested that this finding is due to the transudation of the interstitial edema. It is conceivable that the combination of loss of intratumoral fluid, the mechanical irritation caused by the tumor and the peritoneal inflammation can lead to the production of ascites.[7]

Loizzi et al.[3] described a case of bulky ascites, pelvic mass, associated to bilateral pleural bleeding, with elevation of CA-125 in one 65-year-old patient diagnosed with benign Struma Ovarii.When similar case is treated as one to the described one by Meigs in 1954 (Meigs´ syndrome), they proposed the term pseudo-Meigs´ syndrome for the case. In spite of the origin of the effusions to remain obscure as much in the Meigs' syndrome as much in pseudo-Meigs' syndrome, some mechanisms are still suggested, like active fluids secretion by the tumor or by the peritoneum, by lymphatic and/or veined obstruction, low protein serum level and the presence of toxins and inflammatory products. It is still seen the straight relation between the presence of the tumor and the effusions, having in mind that, when the tumoral mass is moved, quick regression of the effusions takes place.[4]

The levels of CA-125 generally are inside the limits of the normality, in most of the cases of Struma Ovarii, meantime, as well as we turn back in our case report, other authors also found the association of elevated levels of CA-125 and Struma Ovarii. Loizzi et al.[3] had a case report of Struma Ovarii in 83-year-old patient, presenting bulky ascites (approximately three litters) and significant elevation of the levels of CA-125 (1570 u/ml). Some authors hypothesized, in this case, that the elevation of the levels of CA-125 was due to the bulky ascites, in spite of the value were well above what typically takes place in the secondary ascites to the benign ovaries disease.

In a second hypothesis, the inflammatory reaction caused by the ascites or by the tumor can explain the elevations of the levels of CA-125. In the Meigs' syndrome, the immunohistochemistry expression of CA-125 is evident in the surface of the peritoneum of which in the tumoral layer; without associating high levels of CA-125 with ascites volume or with the size of the tumor.

In this described case, there was a value of CA-125 superior to 600 u/ml associated with ascites with a volume of 6 liters.

In spite of the elevation of CA-125 to take place in begin and malignant pathologies, the positive predictive value are 60 % for cancer. There is not a specific biological marker for the diagnosis of Struma Ovarii.[2]

The radiological aspect is specific to Struma Ovarii. In the radiological evaluation, it can present itself as solid, liquid lesions and in its majority, mixed. In the cystic lesions there are septa delimiting cavities with vascularization.[8] Flow is detected through the

Doppler examination, in spite of the limitations of the transabdominal evaluation in this cases.[8] Calcifications can be seen in the ultrasound and confirmed in the tomography, but are ignored in the magnetic nuclear resonance.[8]

Malignant transformation takes place in less than 33 % of the cases.[5] The first description of malignant Struma Ovarii was done by Gottschalk in 1899, and was called malignant folliculoma of the ovary. The most frequent malignant forms are papillary carcinoma or vesicular carcinoma. Some malignant criteria (capsular and/or vascular invasion) are for times difficult to be noticed or insufficient for the diagnosis in the absence of metastasis.[1] The malignant dissemination takes place in only 5% to 6 % of the cases, and it occurs through regional dissemination to pelvis and paraortic lymphonodes, for extension for the epiplon, peritoneal cavity and contralateral ovary, and for hematogenic route for liver, lungs, brain and bones.[2]

The cytology of the ascitic fluid can detect malignant cells in most of the cases, though it still has a significant rate of false negative.[5]

The treatment of benign Struma Ovarii consists of the unilateral ooforectomy without being need for complementary treatment, except for the alteration of thyroid function. In these cases, it is necessary to proceed to the treatment of the dysfunction before the surgery.[2] In the case of malignancy, due to the fact of being quite rare, there is still not a consensus in the literature. It is necessary to considerer more a thyroid than an ovary malignant neoplasia. Being so, it is extolled by some authors that the treatment is similar to the carried out for the malignant neoplasia of the thyroid, with ovarian surgery, thyroidectomy and complementary treatment with Iodine 131.[2] Other authors suggest a conservative treatment with unilateral ooforectomy, when it was reserved to young patients with wish of having children. The total hysterectomy with bilateral ooforectomy would be carried out in cases of extensive or metastatic tumors.[1] The total thyroidectomy, for some authors, is reserved for the metastatic cases or recurrences, and it must be carried out by two reasons: to move the tumor to the treatment with radio-iodine and to remove a metastasis of thyroid câncer.[1]

In spite of the combination of three clinical findings (ascites, pelvic mass and elevation of the CA-125 levels) highly suspected of a malignancy, exist, in rare cases, benign ovarian pathologies, and the Struma Ovarii should make part of the differential diagnosis.

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