

A rare case of trichilemmal ovarian carcinoma. Case report and review of the literature

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SUMMARY: A rare case of trichilemmal ovarian carcinoma. Case report and review of the literature.

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Trichilemmal ovarian carcinoma is very rare.

A 52-years-old woman was admitted to University Hospital Department suffering from pain in the left iliac fossa lasting for three months. Uterus was normal in size, painless as well as right adnexum. On the left site ovary was increased in volume and slightly sore. Transvaginal ultrasounds showed a 97x65x86 mm mixed vascularized unilocular mass.

CT scan detected a pelvic expansion with a heterogeneous density due to the presence of different structures varying from fat to bone tissue. A

proper informed consent was obtained and a suprapubic transversal laparotomy according to Pfannenstiel was carried out.

Ovarian mass and ipsilateral tube were removed with no rupture.

The anatomical extemporaneous result was of benign dermoid cyst.

The postoperative course was uneventful and after three days the patient was discharged in regular conditions and was recommended to come for final anatomical result. Following a month, the final histological answer was of 15 mm trichilemmal malignant tumor in a context of a large benign dermoid cyst. The neoplastic lesion appears to be entirely contained within the limits of the swollen mass. The outer surface was with no lesions.

A case of trichilemmal malignant tumor involving ovary has not been published so far.

KEY WORDS: Trichilemmal tumor - Ovarian cancer - Adnexial cyst - Dermoid cyst - Teratoma.

Introduction

Trichilemmal tumor is a rare cutaneous tumor that usually originates from the bulb of the hair follicle and affects areas of the scalp exposed to the sun. It simulates squamous cell carcinoma with a low degree of skin differentiation (1, 2) and has an unpredictable biological behavior giving rarely distant metastases.

Only a few cases of cutaneous malignant trichilemmal tumor have been reported in the literature so far (3). Trichilemmal cancer can also affect the ovary, as some types of ovarian cancer arise from totipotential cells of dermoid cyst. Generally this sort of tumor is benign.

It is the first case reported in literature of ovarian malignant trichilemmal tumor.

Case report

A 52-years-old woman was admitted to University Hospital Department suffering from pain in the left iliac fossa lasting for three months. At gynecologic bimanual visit, uterus was normal in size painless as well as right adnexum. On the left site ovary was increased in volume and slightly sore.

Transvaginal ultrasounds showed a 97x65x86 mm mixed vascularized unilocular mass, the structure of which was inhomogeneous, containing hairs while edges were regular.

First claimed hypothesis was benign dermoid cyst.

Abdominal CT scan without and with contrast medium detected a pelvic mass with a heterogeneous density due to the presence of different structures varying from fat to bone tissue (Figure 1).

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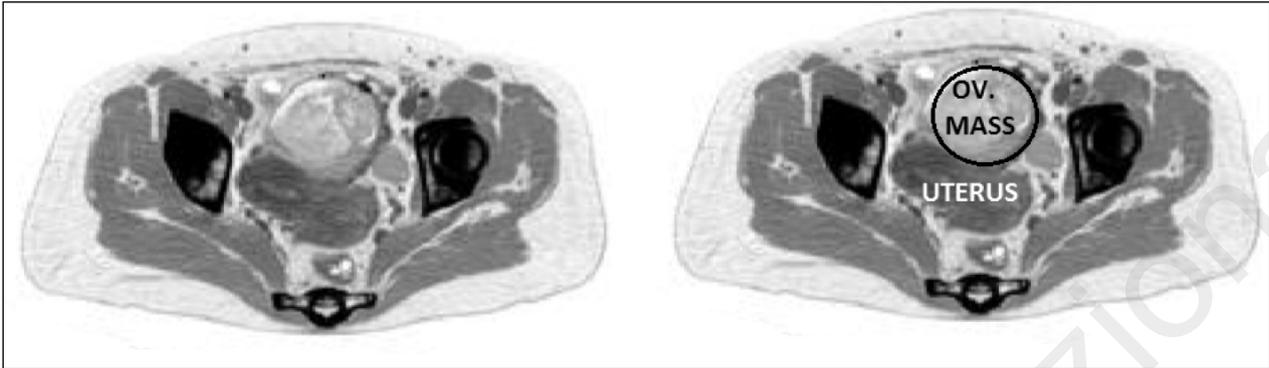


Figure 1 - CT showing left ovarian mass.

The patient underwent surgery. A proper informed consent was obtained and a suprapubic transversal laparotomy according to Pfannenstiel was carried out.

After opening peritoneal cavity, uterus and right ovary appeared of normal and regular shape, whereas the left ovarian was transformed into a swelling 10 cm mass with regular surface.

Clamping, section and ligature of the left infundibulo-pelvic ligament and of the proximal left ipsilateral adnexal peduncle were performed, left adnexum including ovarian mass (Figure 2) was removed without rupture and sent for intraoperative pathological examination. The anatomical result was at the beginning of benign dermoid cyst.

The postoperative course was uneventful. After three days the patient was discharged in regular conditions and was recommended to come for final anatomical result. Following a month, the final histological answer was of 15 mm trichilemmal malignant tumor in a context of a large benign dermoid cyst. The neoplastic lesion appears to be entirely contained within the limits of the swollen mass. The outer surface was with no lesions.

Discussion

The malignant trichilemmal tumor is a rare cutaneous carcinoma that usually arises in exposed areas of the body. It is more common for women and the most frequent sites in which it occurs are scalp, neck, upper chest area and other cutaneous tissues. It rarely develops at other sites (4, 5).

In this case the tumor occurred in a 52-year old woman affecting the left ovary. The imaging aspect

of this tumor was of a vascularized cystic structure with heterogeneous content such as dermoid cyst.

The patient presented only with pain in the left iliac fossa, but the clinical data and imaging techniques were not sufficient to establish the diagnosis that was only obtained later by final histological examination.

The pathogenesis of the trichilemmal malignant tumor remains unknown. With regard to cutaneous trichilemmal tumor, studies have pointed out solar exposure having a significant role in the pathogenesis of the disease, while it is more difficult to establish the pathogenesis for the homologous ovarian malignant tumor (6) (Figure 2).

The clinical evolution of the trichilemmal tumor is unpredictable, in many cases, as regards origin and metastasis.

In rare cases distant metastases have been reported.

Genetic studies have been carried out. DNA was isolated from microdissected areas of proliferating trichilemmal cyst and carcinoma respectively, and PCR-based microsatellite loss of heterozygosity (LOH) analysis as well as p53 gene sequencing performed. LOH analysis showed that the proliferating trichilemmal cyst (PTC) retained chromosome arm 17p (where the p53 gene resides), whereas the carcinoma was associated with the loss of this allele. All the other loci examined were retained including 3p, 9q, 13q and 17q in both tumor parts. A clonal origin of the proliferating trichilemmal cyst and trichilemmal carcinoma was advanced and, moreover, a complete loss of the wild-type p53 was retained responsible for malignant evolution (7).

Close follow-up is essential for the early diagnosis of tumor metastases (8, 9).

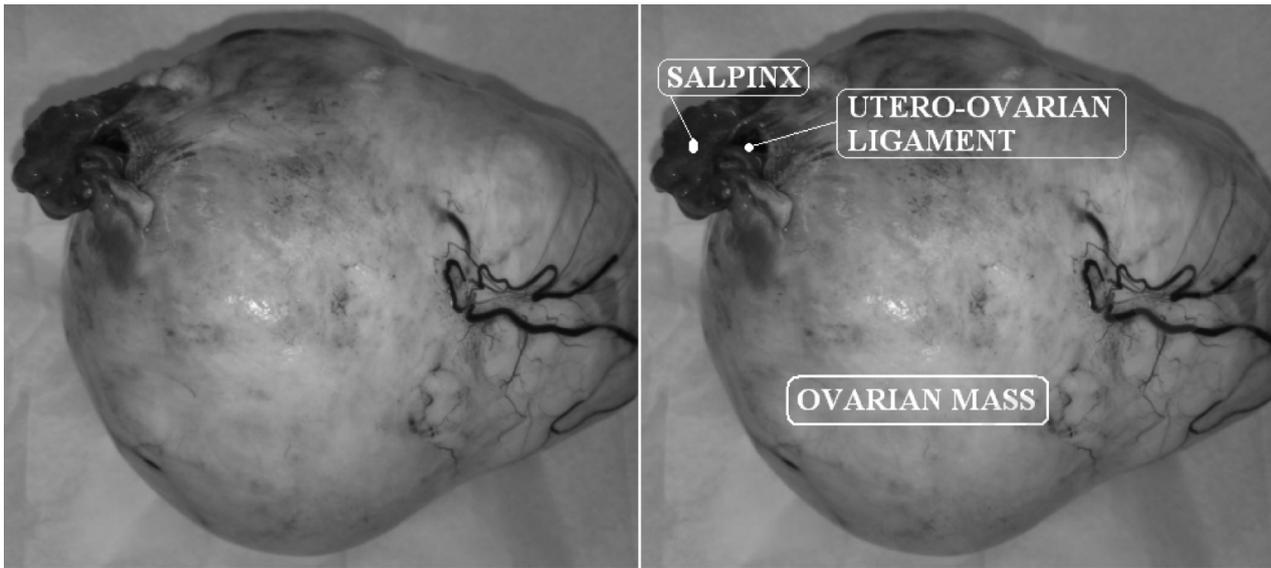


Figure 2 - Left adnexum containing malignant trichilemmal tumor.

Despite the laparoscopy, today, is the method of choice in the treatment of ovarian cyst, as it decreases morbidity and the incidence of postoperative adhesions compared to laparotomy, our choice was to proceed laparotomically. This route allowed to remove the mass with no rupture avoiding spread of neoplastic cells.

Extemporary examination is important for establishing how to end the procedure, however we can't exclude that the final histological exam may give a different result as it happened in our case.

A correct informed consent to the patient is mandatory.

Conclusions

Regarding malignant proliferating of trichilemmal mass, only 39 well-documented cases have been published to date in the international literature and they refer to skin lesions (10). A wide range of trichilemmal cyst with minimal hyperplasia, to full-blown proliferating has been observed. Rarely, patients have ordinary trichilemmal cysts on their scalp associated with malignant mass. Besides to trichilemmal keratinization of hair, proliferating trichilemmal cysts shows a wider combination of differentiation, including features of the follicular in-

fundibulum, the lower nonkeratinizing portion of the follicular outer root sheath and sebaceous cells. Severe inflammatory reaction and cellular atypia, were also observed in proliferating trichilemmal tumor. Lesion similar to pseudocarcinoma, while maintaining its benign biologic behavior has been reported, as well (11). Regarding imaging diagnosis with ultrasounds, the aspect of mass is usually inhomogeneous. At CT trichilemmal tumor may appear as a poorly marginated soft-tissue mass with isointense signal on T1-weighted MR images and hyperintense signal on T2-weighted images. Large areas of high signal intensity caused by necrosis are also found within the tumor on T2-weighted images. After administration of contrast, the mass may show a significant enhancement, with large portions remaining unenhanced. In other cases CT scans may detect a well-encapsulated cystic mass with multiple speckled calcifications in a wall of variable thickness. Several foci of smooth soft-tissue protuberances from the inner wall of the mass, which corresponded histologically to proliferating portions of trichilemmal cyst may result (12-21). The ovarian trichilemmal carcinoma is a very rare event. The preoperative diagnosis is very difficult. The pathogenesis is unknown. The complete removal before arising metastasis is the fundamental step to achieve patient recovery (22-27).

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