Case Report

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When Pelvic Mass Remains a Diagnostic Challenge: A Case Report of a Large Extraovarian Fibrothecoma

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Abstract

Extraovarian sex-cord-stromal tumors, especially fibrothecoma, are a rare entity and therefore lead to frequent diagnostic uncertainties. We report the case of a fibrothecoma originating from the right uterosacral ligament in an 83-year-old female presenting with mixed urinary incontinence due to a tumorous lesion absorbing the entire pelvis. The patient had previously undergone multiple pelvic surgeries including hysterectomy and bilateral adnexectomy. In a preoperative percutaneous biopsy the lesion was diagnosed initially as PEComa. After surgical extraction of the tumor and subsequent thorough histopathological, histochemical and immunohistochemical examination the initial diagnosis had to be changed to fibrothecoma. Diagnosis of extraovarian sex-cord-stromal tumors remains challenging and tight interdisciplinary exchange is necessary to improve diagnostics and consecutive therapy in patients with this rare tumor entity.

Keywords: Extraovarian; Sex-cord-stromal tumor; Fibrothecoma; PEComa; Pelvic mass

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Introduction

Ovarian sex-cord-stromal tumors (SCST) represent approximately 7% of all ovarian tumors [1]. Extraovarian location of a sex-cord-stromal tumor is an absolute rarity, and among these fibrothecoma is even more scarce [2-5]. The exact histogenesis of SCST is not yet thoroughly understood. We report the case of an extraovarian fibrothecoma originating from the uterosacral ligament. To the best of our knowledge this is the first report of an extra-ovarian sex-cord-stromal tumor emerging from that anatomical location. The main goal of this report is to increase awareness of extraovarian SCSTs to improve preoperative diagnosis.

Case Report

We report the case of an 83-year-old Caucasian woman who was admitted to our clinic for further assessment due to a mixed urinary incontinence. She had a vast prior gynecological medical history. Amongst others she had undergone a vaginal hysterectomy in 1979 because of hypermenorrhea and in 2005 a midline longitudinal laparotomy with bilateral adnexectomy, omentectomy and peritoneal biopsy had been performed for a mucinous borderline tumor of the left ovary. In addition, an anterior colporrhaphy with mesh inlay in 2010 as well as a colpoperineoplasty with a subtotal colpocleisis in 2014 had been performed in order to correct a combined recto- and enterocele.

Vaginal examination revealed a mesh erosion in the anterior vaginal compartment, measuring 10 × 10 mm, without any sign for infection. Vaginal ultrasonography showed a large hypoechogenic pelvic mass with slight focal doppler enhancement. In the computed tomography a solid intraperitoneal 12 × 13 × 12 cm process with compression of the bladder and rectosigmoid colon and slight concomittant ascites were seen. A suspicious hypermetabolic activity was observed at the PET scan in two distinct locations, one in the pelvic lesion and a second one localized in the right lower lobe of the thyroid. These findings were suggestive of malignancy. Subsequent clinical, laboratory and ultrasonographic assessment showed no suspect lesions in the thyroid. Histopathological examination of a transabdominal biopsy of the pelvic mass showed a neoplasia comprising fusiform cells most likely interpreted as a perivascular epitheloid cell tumor (PEComa) (**Figures 1-3**).

We performed a re-laparotomy with extensive adhesiolysis, the

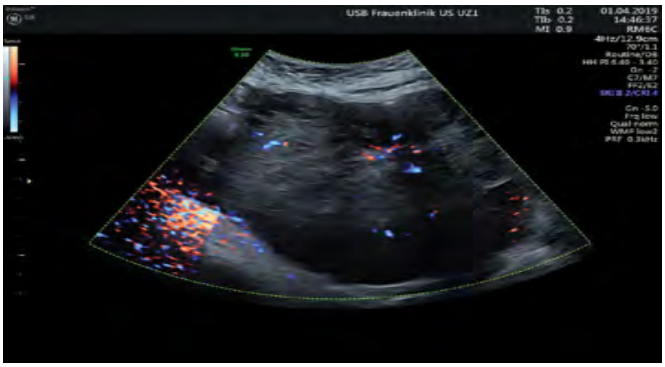

Figure 1 Transvaginal sonography with slight focal Doppler enhancement.

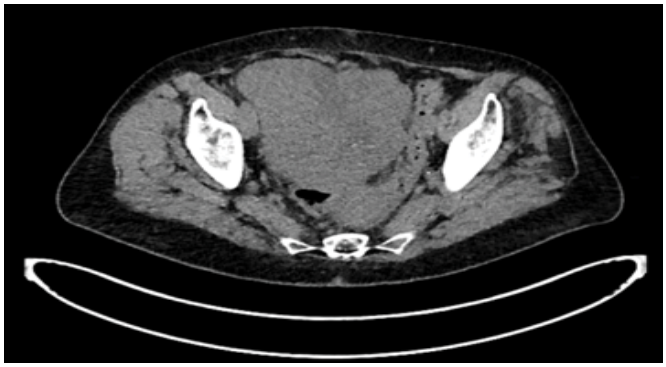

Figure 2 CT scan showing the pelvic lesion with slight surrounding ascites.

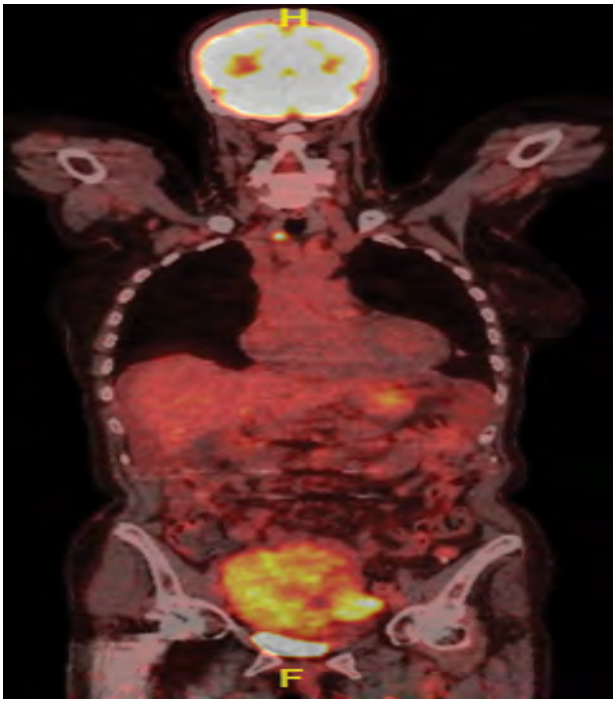

Figure 3 PET scan with FDG accumulation in pelvic mass.

pelvic mass was found to originate from the right ligametum sacrouterinum. The frozen section showed a tumor of fusiform cells without any signs for malignancy. The tumor was completely resected, the staging showed no further macroscopic lesions, and the vaginal mesh erosion was closed after partial resection of the mesh graft. The postoperative course was uneventful. There was no evidence of recurrence within a nine-month postoperative period (**Figures 4-6)**.

Histopathological examination

On gross examination, the multilobular firm tumor had a slightly yellowish cut surface. Histologically, the fusiform tumor cells were arranged in cellular bundles. The tumor cells contained oval to spindled nuclei. Proliferative activity was very low, with <1

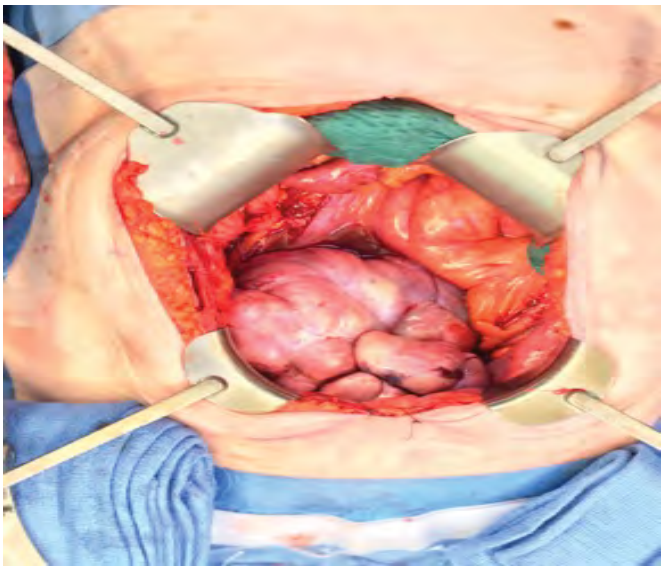

Figure 4 Intraoperative situs.

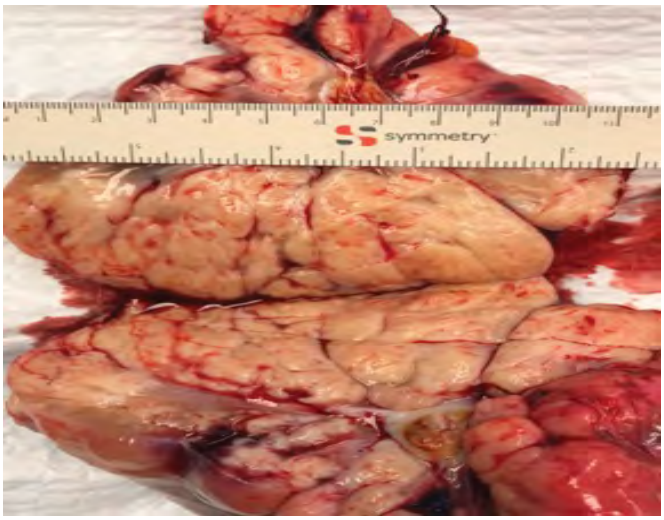

Figure 5 Excision in toto.

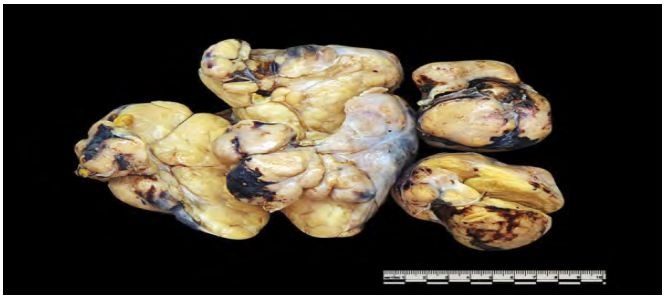

Figure 6 Striking yellowish pigmentation.

mitosis per 10 HPF (high-power field). In hotspots the maximum Ki-67 proliferation index was 5%. In 20 representative tissue blocks, no cytological atypia or transformation into a sarcoma could be detected. For further characterization of the tumor, numerous histochemical and immunohistochemical analyses had been performed.

On immunohistochemistry the fibroma cells were positive in 100% for Wilm's tumor gene (WT 1) and CD56. In addition, tumor cells expressed the melanocytic marker Melan-A (80%), which had been the cause of the misdiagnosis as a PEComa in the preoperative biopsy, alpha smooth muscle actin (SMA, 50%) and inhibin (40%). Slight positivity was found for Desmin (10%). The tumor cells were negative for pan-cytokeratin 22, CD10 and CD34 as well as for Calretinin. Decisive for the differentiation from the initially diagnosed PEComa was the typical finely woven fiber network around single tumor cells or small groups of tumor cells in the Novotny reticulin fibre staining.

The final pathological diagnosis was of a benign sex cord-stromal tumor belonging to the group of thecoma-fibroma arising from the ligamentum sacrouterinum (**Figures 7-10).**

Discussion

The pre-operative diagnosis of extraovarian SCSTs remains challenging due to non-specific or misleading characteristics on small preoperative biopsies, as well as the rarity of this entity in a sometimes very unusual location. Modern imaging modalities such us ultrasound, CT-scan and MRI are helpful but no specific features suggest the preoperative diagnosis of extraovarian SCST [6]. Pelvic MRI was not performed in our case, but to exclude metastases in the light of prior medical history, we performed a PET scan.

In the initial biopsy, a PEComa was diagnosed based on morphological features and the immunophenotype typical of this entity (SMA and Melan-A positivity).

Reviewing actual literature PEComas of the gynecological tract are rare tumors which were first recognized and described twenty years ago. Histologically, PEComa is characterized by the presence of predominantly epithelioid cells with clear, granular or eosinophilic cytoplasm, arranged in nests or sheets, with little intervening stroma. PEComas are defined by the immunohistochemical (IHC) co-expression of myoid markers

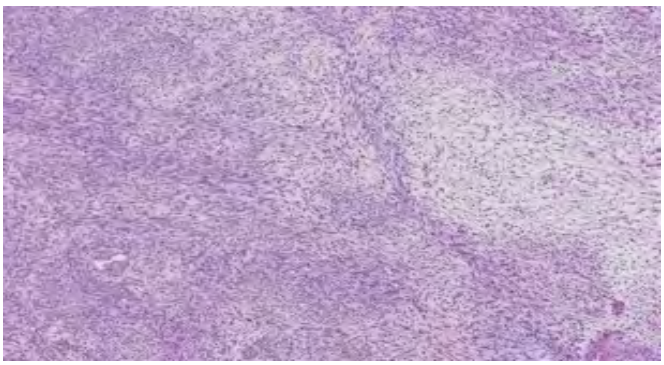

Figure 7 HE: 100x: HE fusiform cells with fascicular growth pattern and brighter areas with less cellularity.

Figure 8 WT1 100x: WT1 diffusely positive.

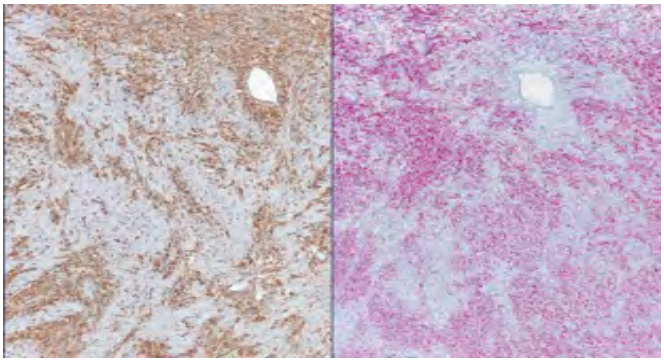

Figure 9 SMA-MelanA × 100b: SMA left, Melan A right from the same tumor section with inverse staining pattern.

(SMA, desmin, caldesmon) and melanocytic markers (HMB-45, Melan-A, MiTF) [7]. To our knowledge no PEComas have been reported originating from the uterosacral ligament.

Melan-A is not only produced in melanocytic tumors and PEComas but also in steroid hormone producing cells of the adrenal cortex or of the gonads [8,9]. The decisive additional analysis for the differentiation of the two entities in our case, was the histochemical silver staining (Novotny) depicting the characteristic finely woven fiber network around single tumor cells or small groups of tumor cells of SCST which is not present

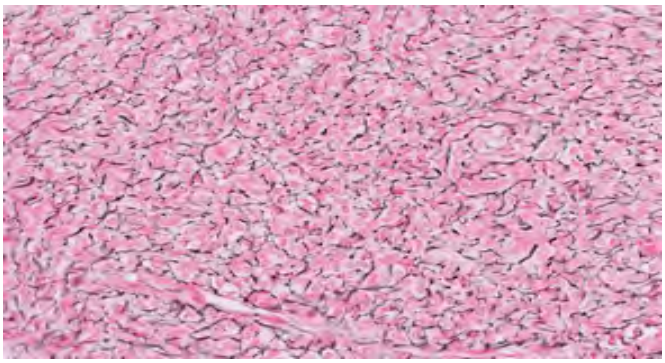

Figure 10 Novotny-400x: Novotny reticulin fiber staining with typical finely woven fiber network around single tumor cells or small groups of tumor cells characteristic for fibrothecoma.

in PEComas.

Extraovarian sex cord-stromal tumor (SCST) is an extremely rare entity only described in individual case reports and reviews [5,10]. The etiology for the development of SCSTs in an extraovarian location is unknown and its histogenesis remains unclear. It is hypothesized that such tumors originate in postsurgical and postinflammatory implantations, an accessory or supernumerary ovary or as a result of ovarian autoamputation [11,12]. Strikingly most reported cases had a history of prior pelvic surgery which favors the theory of postsurgical implantation. Our patient had a history of extensive prior pelvic surgery supporting the hypothesis of postoperative implantation as a possible reason for the development of the extraovarian fibrothecomas.

Even though extraovarian SCST have been described as benign, estrogen production by these tumors can lead to endometrial carcinoma [13,14]. Further these tumors can mimic metastatic ovarian cancer with elevated levels of cancer antigen 125 (CA-125) or manifest as Meigs' syndrome, described as the triad of ascites, unilateral hydrothorax and benign ovarian tumor [15]. Current tumor markers are not reliable in diagnosing SCST.

Intraoperative fresh frozen section is helpful to prevent excessive surgery. However for definite pathological diagnosis thorough immunohistochemical und histochemical testing is indispensable as our case has exemplarily shown. Diagnoses made on preoperative punch biopsies may be wrong because the tissue is not representative for the entire tumor. Furthermore particular diagnostic is often not considered for very rare entities or in case of unusual anatomical localization. It is therefore important to constantly question existing diagnoses in the light of new findings in order to avoid false diagnoses and enable optimal therapy.

Conclusion

Clinicians, radiologists and pathologists should consider the diagnosis of extraovarian SCST in women presenting with oddly shaped mass in the pelvis with normal ovaries or status post bilateral oophorectomy. Tight-knit interdisciplinary exchange is necessary to improve diagnostics especially in consideration of lack of clinical data due to the scarce occurrence of this tumor entity.

Conflict of Interests

The authors declare no potential conflicts of interest.

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