

## A RARE CASE OF ABERRANT MIGRATION OF PRIMORDIAL GERM CELLS - UTERINE YOLK SAC TUMOR

L. Păiușan, C. Ülgüt, P. Deme

**Abstract.** Yolk sac tumor of uterus is a very rare tumor that develops in the uterus as a result of aberrant migration of primordial germ cells. We present a case of 57 years old woman in climax, which presents at gynecological examination after vaginal bleedings. Biopsy report suggested non-keratinized epidermoid carcinoma microcellular with areas of clear cell carcinoma, poorly differentiated G3, with areas of infarction and acute inflammation associated. The mass was excised and sent to our pathology department for histopathological examination. We reported Yolk sac tumor of uterus with hepatoid pattern T2aN0M0. The section from the uterine cervix and ovary did not show any remarkable pathology.

### Introduction

Yolk sac tumor, classified as germ cell tumor, is a rare malignant tumor of cells that separates the yolk sac of the embryo. Physiologically, from these cells develop ovaries/testes. The cause of migration of these germ cells at various levels and their transformation into malignant cells is not known. Their malignant transformation is 10% of all germ cell cancers.

Most yolk sac tumors of the female genitalia is localized in the ovaries. Rare cases have been however reported on the vulva, cervix and endometrium. Yolk sac tumors located in the pelvis outside the ovary, as our case, are unusual. It is believed that these tumors occur in the womb as a result of aberrant migration of primordial germ cells.

### Case report

We present the case of a female patient, aged 57 years, in climax, which presents for gynecological bleeding. It is practiced bioptic uterine curettage showing on histopathology, a nonkeratinised microcellular epidermoid carcinoma with areas of clear cell carcinoma, poorly differentiated G3, with areas of infarction and acute inflammation associated. It is decided surgery uterus removal with tumor mass. During surgery it is found an increased volume of the uterus with multiple intramural and subserosal nodules, and is practiced total hysterectomy with bilateral ovariectomy and

surgical specimen is sent for histopathological examination.

### Macroscopic examination

Surgical specimen consisting of uterus and annexes was examined both macroscopically and microscopically. On macroscopic examination uterus size approx. 9x5 cm, external surface was nodular with multiple intramural and subserosal fibroid nodules of approx. 1,5x2 cm. On the section is observed in the endometrium and myometrium, posterior uterine wall, an infiltrative growing tumor mass with extensive necrosis and areas of hemorrhage.

Ovaries of normal size.

### Microscopic examination

All sections from the uterus showed the morphology of yolk sac tumor - cells with eosinophilic uniform cytoplasm, hyperchromic atypical nuclei, arranged in a hepatoid pattern, constituting a considerable resemblance to hepatocytes.

Characteristic for a yolk sac tumor is the presence of extracellular hyaline droplets and Schiller-Duval body, glomerular-like structures, comprising of a conjunctive-vascular shaft limited by a layer of embryonic tumor cells. Also are presents areas that mimic clear cell carcinoma, extensive areas of necrosis, acute inflammation and hemorrhage.

These changes in tumor infiltrating the endometrium and half superficial myometrium with a poorly differentiated tumor grading.

On the sections in the uterine body is found to be present yolk sac tumor. On the sections from the cervix and annexes are not found neoplastic infiltration or significant histopathological lesions.

Diagnosis: Uterine yolk sac tumor–T2aN0M0

T2a – tumor invades the superficial half of myometrium

N0 – without neoplastic infiltration on regional lymph nodes

M0 – without away metastases

## Discussion

In the literature have been reported only 4 cases of yolk sac tumor in the uterus. Three of them in patients under 12 years and one case of a patient aged 42 years. All 4 patients were treated by total hysterectomy followed by adjuvant multiagent chemotherapy. Two of the patients are surviving long term, and the other two died. Yolk sac tumor, also called endodermal sinus tumor due to its histology similarities with extra embryonic structures of the early embryo, contains a variety of histologic patterns that differ considerably from one to another, however, different patterns can be observed in the same tumor one or two predominant.

Yolk sac tumor may present pattern: microcystic, endodermal sinus, solid, alveolar-glandular yolk polyvesicular, myxomatous, papillary, macrochistic, hepatoid and glandular or endodermal primitive.

Hepatoid pattern, as in the case shown, has a tendency to develop a pure form unassociated with other elements of yolk sac tumor, forming a yolk sac tumor with one histological pattern. It is composed of cells with eosinophilic cytoplasm, and provides a uniform or granular solid pattern having a structure substantially similar to that of hepatocytes.

Yolk sac tumors with such a pattern were called by Teilmann yolk sac tumor with hepatoid pattern.

The presence of Schiller-Duval bodies can be considered pathognomonic for yolk sac tumor, as well as the presence of hyaline droplet eosinophilic both intracellular and extracellular, in HE staining.

These elements, together with the rarity of tumors may cause a problem of diagnosis. This uterine mass composed of hepatocyte-like cells surrounded by connective tissue and the formation of solid aggregates, cords or nests associated with elevated serum AFP levels favor the diagnosis of yolk sac tumor.

Increased serum level of AFP and demonstration of AFP presence in tumor tissue by immunohistochemistry confirmed the diagnosis of yolk sac tumor.

## References:

1. Blaunstein's Pathology of the Female Genital Tract, 5th Edition, Kurman J. Robert, Springer, New-York, 2001
2. Sternberg's Diagnostic Surgical Pathology, 4th Edition, Stacey E. Mills, Carter J. Greenson, 2004
3. Rosai and Ackerman's surgical pathology, 4th Edition, vol-2, 2004
4. Spatz A., Bouron D., et al – „Primary yolk sac tumor of the endometrium: a case report and review of the literature” *Gynecologic Oncology*
5. Joseph MG, Fellows FG, Hearn (1990) Primary endodermal sinus tumor of the endometrium. A clinicopathologic, immunocytochemical and ultrastructural study. *Cancer* (Philadelphia)