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PRIMARY OVARIAN SYNOVIAL SARCOMA — A CASE REPORT

*A. Yordanov[1, *], K. Janeva[2], S. Mateva[3], Y. Kornovski[4], S. Kostov[4], S. Slavchev[4], Y. Ivanova[4], M. Nikolova[3]*

[1]*Department of Gynecologic Oncology, Medical University Pleven, Pleven 5800, Bulgaria*
[2]*Department of Gynecology, Saint Marina University Hospital, Pleven 5800, Bulgaria*
[3]*Department of Pathology Laboratory, Saint Marina University Hospital, Pleven 5800, Bulgaria*
[4]*Department of Obstetrics and Gynecology, University Hospital "Saint Anna", Medical University of Varna "Prof. Dr. Paraskev Stoyanov", Varna 9002, Bulgaria*

Synovial sarcoma is an aggressive soft tissue sarcoma, which most often occurs in the limbs near the joints. It accounts for 5–10% of all soft tissue sarcomas. It extremely rarely affects the pelvis. So far, only 4 cases of primary involvement of the adnexa have been described. We present a case of a 77-year-old female patient diagnosed with a rapidly growing pelvic formation, subsequently diagnosed as monophasic synovial sarcoma of the ovary. Synovial sarcoma derived from the adnexa is a rare disease that is virtually unknown. The diagnosis is complex, and there is a poor prognosis.

***Key Words*: synovial sarcoma, ovary, prognosis.**

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Synovial sarcoma (SS) is an aggressive soft tissue sarcoma (STS) [1]. It most often occurs on the limbs near the joints. It accounts for 5–10% of all STS, making it the 4th most common of this group [2]. It was first described by Simonin in 1865 and was named SS by Sabrazesetal in 1934 [3].

According to the 4th edition of the WHO classification of tumors of soft tissue and bones (2013), SS belongs to the tumors of uncertain differentiation. It is subclassified into two subtypes consisting of a biphasic (BPSS) and monophasic spindle cell (MPSS) type [4]. It extremely rarely affects the pelvis, and so far, only 4 cases of primary involvement of the adnexa have been described [5].

We present a case of a 77-year-old patient diagnosed with a rapidly growing pelvic formation, subsequently diagnosed as monophasic SS of the ovary.

The patient was admitted to the clinic for treatment based on an established formation in the pelvis by ultrasound examination and abdominal pain for 2 weeks. The medical history showed a 25-year history of amenorrhea, 5 pregnancies and 2 children born, and arterial hypertension. The gynecological examination revealed a painful formation in the left ovary with dimensions of 9/5 cm. The ultrasound examination visualized an oval, hypoechoic formation to the left of the uterus with dimensions of 70/50 mm suspected of subserous myoma.

All the laboratory tests showed normal results except for HGB 103 g/L and Urea 12.2 mmol/L. Magnetic resonance imaging of the pelvic floor revealed the following result: formation in the pelvis, on the left, close to the uterus, corresponding to a large, well differentiated, sharply and smoothly outlined myoma with benign character and heterogeneous signal intensity, with dimensions 100/40 mm. Presence of simple cysts in the right ovary; left ovary — not visualized. Bladder — without complaints. Rectum — without complaints. There was no evidence of enlarged lymph nodes. The X-ray of the lungs showed bronchitis changes and aortosclerosis. Based on the tests, we assumed a myoma and decided to perform a total open hysterectomy with bilateral salpingo-oophorectomy (BSO).

Intraoperative finding: uterus — anteverted, with normal shape and size. Left adnexa: ovary — consumed by a tumor formation with a diameter of 7 cm; fallopian tube — of normal length and structure. Right adnexa: ovary — cystic formation with a diameter of 3 cm; fallopian tube — of normal length and structure. Liver — intact. The remaining organs of the abdominal cavity accessible for inspection and omentum are without features. Paraaortic and pelvic lymph nodes — palpably not enlarged.

A total hysterectomy with BSO and biopsy of omentum was performed.

The histological examination of the formation in the left ovary revealed a relatively well-circumscribed nodule composed of highly atypical spindle cells arranged in interlacing fascicles. Numerous atypical mitotic figures were present (Figure, *a,* indicated by arrows) and foci of necrosis (Figure, *b*). Immunohistochemical examination showed positivity for CD99 (Figure, *c*) and epithelial membrane antigen (EMA) (Figure, *d*) and negativity for smooth muscle actin (SMA), desmin, MyoD1, calretinin, WT1 and S-100.

All other gynecological organs removed had normal structure. Based on histological and immunohistochemical findings, we made the diagnosis of MPSS.

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*Correspondence: E-mail: angel.jordanov@gmail.com
Abbreviations used: BPSS – biphasic spindle cell type of synovial sarcoma; BSO – bilateral salpingo-oophorectomy; EMA – epithelial membrane antigen; MPSS – monophasic spindle cell type of synovial sarcoma; SMA – smooth muscle actin; SS – synovial sarcoma; STS – soft tissue sarcoma.

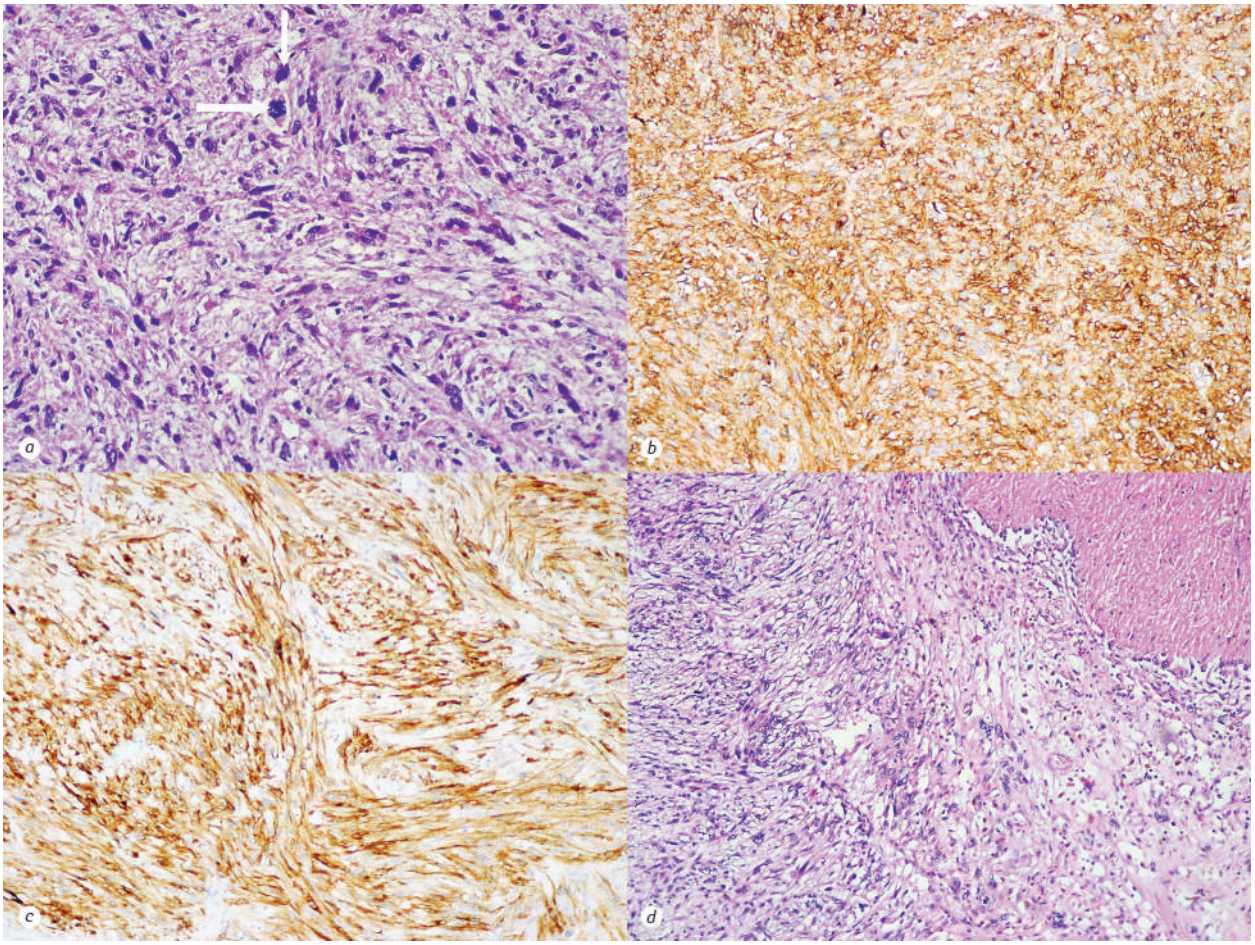

Figure. Histological findings: *a* — numerous atypical mitotic figures; *b* — foci of necrosis; *c* — immunohistochemical examination showed positivity for CD99; *d* — immunohistochemical examination showed positivity for EMA

The patient's postoperative period was uneventful, and the patient was discharged on day 7 without complaints.

The patient was referred for chemotherapy, which she refused. She was lost for follow-up, and based on data from the National Cancer Registry, we found that the patient had died of the disease 16 months later.

SS often occurs in the soft tissues around the larger joints but does not originate from synovial cells [6]. Unlike other sarcomas, SS occurs mainly in younger patients with a mean age of 35 [7]. It is most common in the limbs — about 70% of all cases [8], and these patients have a significantly better prognosis than those with other localizations [9]. In cases where the process is only local, depending on the size and location of the tumor, the 10-year survival rate varies from 8 to 88% [10]. Survival decreases sharply in metastatic disease [11]. SS usually develops slowly but often develops late metastases — 50% of all cases [8], with 80% affecting the lungs [11]. SS can metastasize lymphogenically, with lymph node metastases in 15–20% of the newly diagnosed cases [12]. The standard treatment for SS is tumor resection, often followed by radiation therapy and/or chemotherapy [1].

SS is very rare in the pelvis, and there are only 5 cases of SS starting its development from the adnexa described in the literature (Table).

No conclusions can be drawn about the course of SS from the available data in the literature. Notably, most patients are significantly older than the average for this disease. In two cases, there was a recurrence of the disease. In one of the cases, the patient

Table. Clinicopathological features of primary ovarian and fallopian tube synovial sarcoma described in the literature till now

Case	Age	Location	Size (cm)	Histological subtype	Treatment	Follow-up
Smith *et al.* [13]	90	Left ovary	15	MPSS	Salpingo-oophorectomy	N/A
Mitsuhashi *et al.* [14]	23	Right fallopian tube	15	BPSS	Salpingectomy + 4 cycles chemotherapy (ifosfamide, cisplatin, doxorubicin)	12 months after – local recidive: 3 cycles chemotherapy + surgery + 3 cycles chemotherapy
Nasiri *et al.* (15]	50	Right ovary	15	BPSS	Oophorectomy + 6 cycles chemotherapy	3 years –pulmonary metastasis
Lenz *et al.* [16]	75	Right adnexa, omentum, liver	N/A	BPSS	BSO + omentectomy	Death on the next day
Rahman *et al.* [17]	38	Right ovary	17	MPSS	Salpingo-oophorectomy	Disease-free 27 months after surgery
Present case	77	Left ovary	7	MPSS	TAH+BSO+ omental biopsy	16 months – death

Notes: TAH – total abdominal hysterectomy.

was 6 months without evidence of progression after therapy [14]. In the other case, the patient was stable 5 months later despite refusing treatment [15]. In one case, despite refusing radical surgical treatment and subsequent chemotherapy and/or radiation therapy, the patient had no evidence of recurrence 27 months later [17].

The monophasic variant is more difficult to recognize due to overlapping appearances with other spindle and round-cell sarcomas, and the histological diagnosis is a real challenge in cases with an unusual location like the ovaries. In our case, the histological finding was similar to that of a leiomyosarcoma, but muscle differentiation was excluded because of an adverse reaction with SMA, desmin and MyoD1. Negative calretinin and WT1 staining excluded sex cord-stromal tumor, and negative S-100 showed an absence of nerve sheath differentiation. The diagnosis of MPSS was made based on the histological findings, supported by the positivity of CD99 and EMA.

In our case, despite the fact that after the surgical treatment there was no evidence of persistence of the disease, the patient was offered adjuvant chemotherapy, which was refused. Unfortunately, 16 months later, she died of the underlying disease.

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ПЕРВИННА СИНОВІАЛЬНА САРКОМА ЯЄЧНИКА — КЛІНІЧНИЙ ВИПАДОК

А. Йорданов[1], К. Янева[2], С. Матева[3], І. Корновськи[4], С. Костов[4], С. Славчев[4], І. Іванова[4], М. Ніколова[3]

[1]Відділення онкогінекології медичного університету м. Плевен, Болгарія

[2]Гінекологічне відділення університетського госпіталю "Сент Маріна", Плевен, Болгарія

[3]Патоморфологічний відділ університетського госпіталю "Сент Маріна", Плевен, Болгарія

[4]Відділення акушерства і гінекології університетського госпіталю "Сент Анна", Медичний університет м. Варна, Болгарія

Синовіальна саркома є агресивною саркомою м'яких тканин, яка часто виникає в кінцівках поблизу суглобів. На її частку припадає 5–10% усіх сарком м'яких тканин. Вкрай рідко вражаються тазові органи. До цього часу в літературі описано всього 4 випадки первинної синовіальної сарком придатків. Ми представляємо випадок новоутворення, діагностованого у 77-річної жінки в ділянці малого таза, яке характеризувалося швидким ростом, і згодом було діагностовано як монофазна синовіальна саркома яєчника. Про таку локалізацію синовіальної саркоми дотепер практично не повідомлялося. Діагностика становить певні труднощі, а прогноз — несприятливий.

Ключові слова: синовіальна саркома, яєчник, прогноз.

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