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KLIPPEL - TRENAUNAY SYNDROME IN COMBINATION WITH OVARIAN CANCER

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Abstract. The article presents a clinical case of ovarian cancer, which was diagnosed in a patient with Klippel-Trenaunay syndrome. The clinical picture of the disease was dominated by pain and an increase in the size of the abdomen, urinary retention and stool, severe general weakness, malaise, and palpitations. After a diagnostic examination and preliminary preparation, the patient was operated on for health reasons in a hospital at the oncogynecology department of the Tashkent City branch of the Republican Specialized Scientific and Practical Medical Center of Oncology and Radiology with a preliminary diagnosis of an ovarian tumor. Interrooperatively, revealed angiodysplastic changes in the vessels of the internal organs, and the nature of the ovarian tumor, G2 adenocarcinoma, was also clarified. This clinical case is a unique example of a combination of congenital angiodysplasia syndrome and ovarian cancer. These data may help in the management of patients with this pathology and improve the survival rate of patients with Klippel-Trenaunay syndrome.

Key words: Klippel-Trenaunay syndrome, ovarian cancer, angiodysplasia, express histology.

Introduction. Klippel-Trenaunay syndrome is a congenital malformation of the venous system, which is characterized by a violation of the anatomy of the veins of the lower extremities. Sometimes you can find another name for this disease - dysplasia of the main veins. A common characteristic of this condition is abnormal development of the blood vessels, skin, muscles, and bones. Based on this, doctors

distinguish three main signs by which the presence of pathology can be determined: 1. angiomas in the form of blue nevi, which appear even at birth, less often - in childhood. Often referred to as port-wine stains, they are caused by swelling of small blood vessels near the surface of the skin. For angiomas, in the context of the Klippel-Trenaunay syndrome, clear boundaries and color are characteristic - from pale pink to maroon; 2. hypertrophy - a visually distinguishable increase in the affected limb. A pathological change in size affects the volume of active movements, can cause a feeling of heaviness and pain; 3. varicose lesion of the lower limb, which manifests itself in childhood and adolescence [1].

Klippel-Trenaunay syndrome occurs in people who do not have this pathology in their family, and is not inherited. This condition is a consequence of gene mutations that occur even in the early stages of fetal development in the womb. The exact cause of Klippel-Trenaunay syndrome has not yet been discovered, but there are several assumptions. It is believed that a number of teratogenic factors can affect the development of the embryo through damage to the embryonic vascular epithelium and cause inevitable changes. These include: taking drugs in the first and second trimesters that have a teratogenic effect; radiation exposure; infectious diseases during pregnancy, for example – toxoplasmosis [2,5].

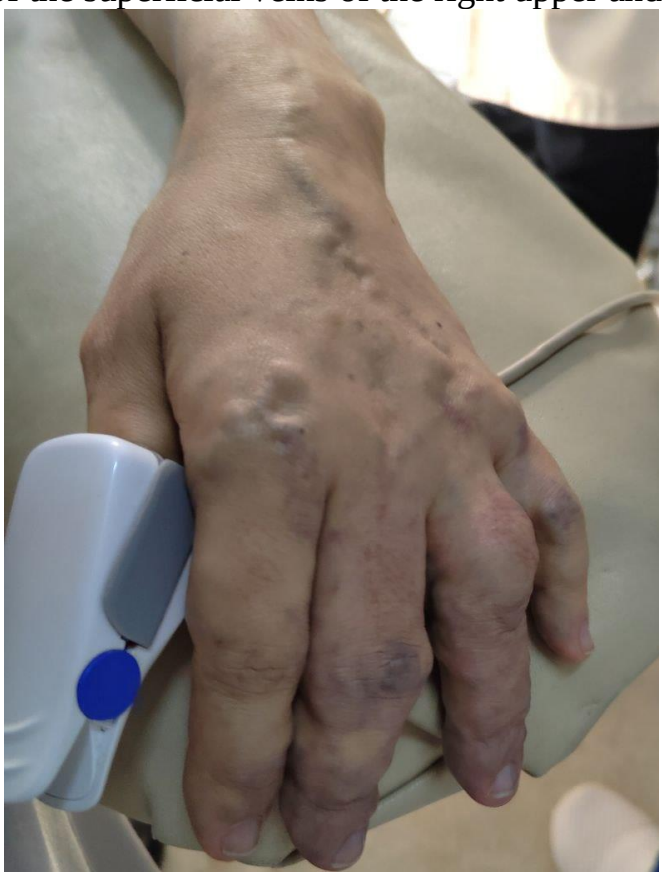
The anomaly is diagnosed immediately at birth, since angiomas already appear in the form of a nevus or wine stains. The next symptom that parents later notice is an increase in the affected limb, which is the result of abundant blood supply. Visually, the difference between the affected limb and the healthy one is very clear. The bones are elongated and thickened, the leg becomes much larger in girth. Varicose veins are often complicated by thrombophlebitis, phlebothrombosis and lipodermatosclerosis [3,4,7]. There are three main ways to diagnose varicose disorders in Klippel-Trenaunay syndrome: duplex scanning of vessels of the lower extremity in order to visualize blood clots, valves and vein anatomy in a patient; MRI and/or CT help to visualize the anatomical structure of the veins and detect the presence of blood clots in locations that are inaccessible with duplex scanning. This method is also effective in assessing the condition of bone tissue and joints; Phlebography is used to identify anatomical features and the presence of arteriovenous fistulas [5,6,8].

Due to the rarity of this pathology, we present a clinical case that may be of particular interest to surgical doctors.

Patient M., 35 years old, was admitted to the department of oncogynecology of the Tashkent city branch of the Republican Specialized Scientific and Practical Medical Center of Oncology and Radiology with complaints of pain and an increase in the size of the abdomen, urinary retention and stool, severe general weakness, malaise, palpitations, dizziness. Before hospitalization, the patient was examined: MRI of the abdominal cavity dated 11/01/22: Cystic formations are determined in the abdominal cavity: 1. Mesogastric region with dimensions of 21x11x14cm, cystic-solid structure. 2. Adjacent to the lower part of the formation described above, a cystic-solid formation is determined with dimensions of 15.6x13.1x10.1 cm. 3. A similar formation is determined on the left surface of the formation described above with dimensions of 9.2x15.3x10.4 cm without visible signs of infiltration of the surrounding organs.

Conclusion: MRI signs of cystic formations of the abdominal cavity, possibly outgoing right ovary. Cystic formations in the liver and spleen, along the right ovarian vein. Tumor markers CA-125: 106 U/ml, HE4: 492.7 pmol/l, ROMA index 95.47%. Biochemistry of blood from 11/01/22: creatinine in the blood 0.092 mmol/l, urea 10.4 mmol/l. Recurrent venography of the lower extremities 01.11.22: MSCT signs of varicose veins of the superficial and perforating veins of the right lower extremities. Indirect signs of the presence of arteriovenous fistulas at the level of n / 3 of the lower leg and foot. An increase in the volume of soft tissues of the right lower extremities. Consultation of a vascular surgeon dated 11/01/22: the first stage of treatment was recommended to the patient, surgical intervention for cystic formation of the abdominal cavity. Ultrasound 03.11.22: echocardiography of tumor ovari, adenomyosis, endometriosis of the cervix. Liver cyst.

From the anamnesis since childhood, she suffers from congenital angiodysplasia of the right leg (Klippel Trionene Syndrome), for which she was operated on in 1998 in a Moscow hospital. Locally, the right leg is visually hypertrophied, local thermoregulation is disturbed, there are periodic aching pains and varicose deformity of the superficial veins of the right upper and lower extremities.



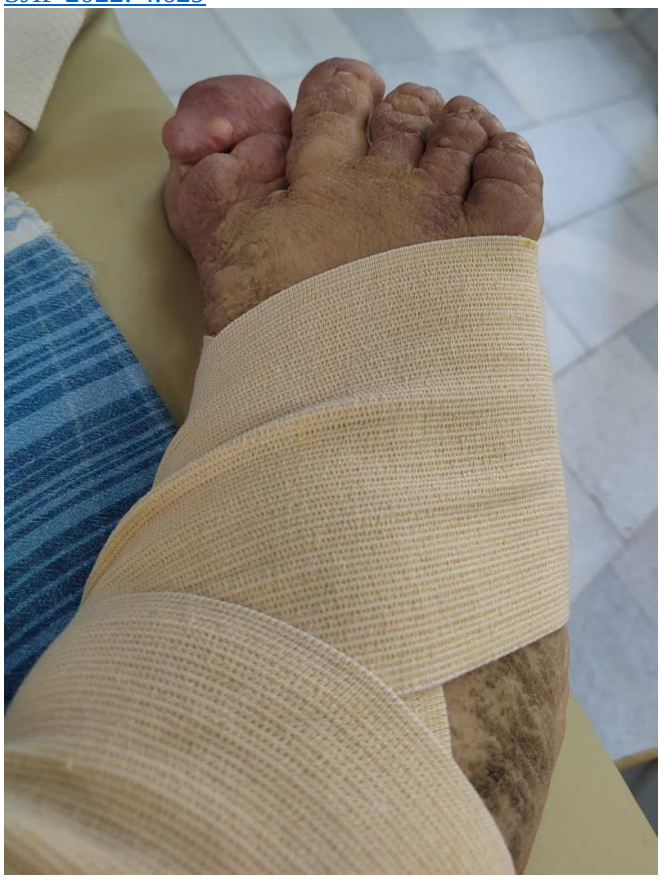


Figure 1. View of the patient's right arm and leg.

Status Genitalis: external genitalia are asymmetrically developed, the right labia is edematous, (due to varicose veins) female-type hair. In the mirrors: the cervix is clean, the discharge is light. Per Vaginum: the entire abdominal cavity is replaced by a tumor originating from the small pelvis, densely elastic consistency, fixed, limited mobility, painful on palpation, the body of the uterus and appendages are not palpated separately. Parameters are free. The patient received preoperative preparation. Consulted and examined by a cardiologist, angiosurgeon, therapist, anesthetist. 05.11.2022 was performed an organ-saving surgical intervention according to vital indications, with an extremely high risk of thromboembolic complications, in the amount of "LAPAROTOMY. REVISION. RIGHT-SIDE TUBOVARECTOMY. RESECTION OF THE LEFT OVARIAN. Biopsy of the peritoneum and greater omentum.

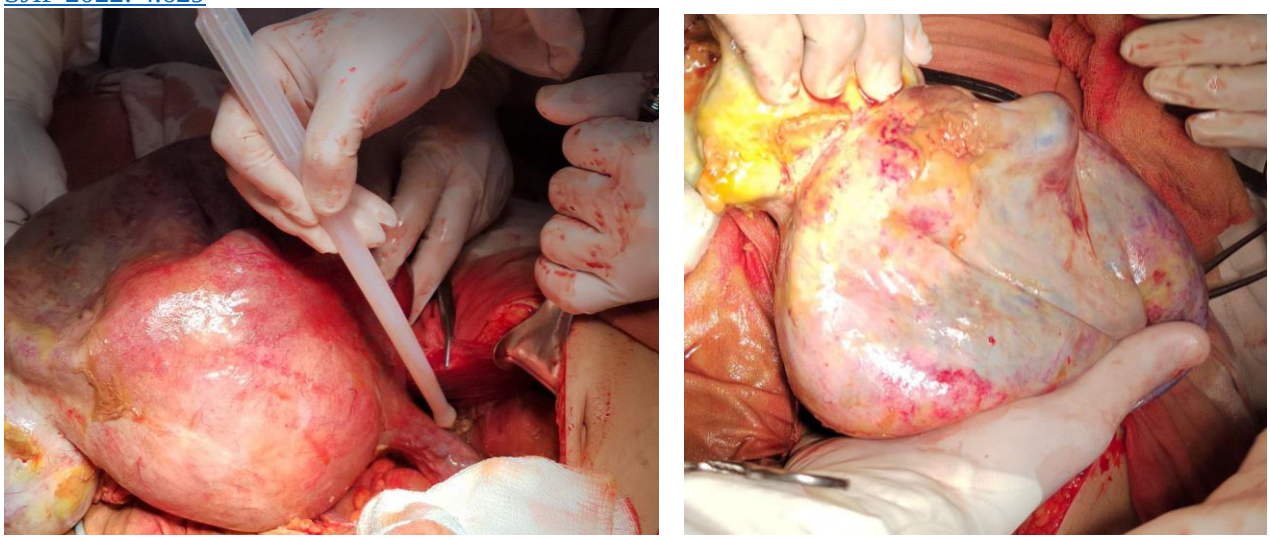


Figure 2. Intraoperative picture of an ovarian tumor.

Histological analysis 105473/22 dated 11/14/22: Adenocarcinoma G-2 with areas of squamous differentiation, endometrioid cyst of the right ovary. Endometriosis of the right fallopian tube. Multiple serous and mucinous cystadenomas of the left ovary. Inflammatory infiltration of the peritoneum. Seal without features.

In the postoperative period, the patient received antibacterial, restorative, anticoagulant, cardiotropic, analgesic therapy.

Figure 3. Resected giant ovarian tumor.

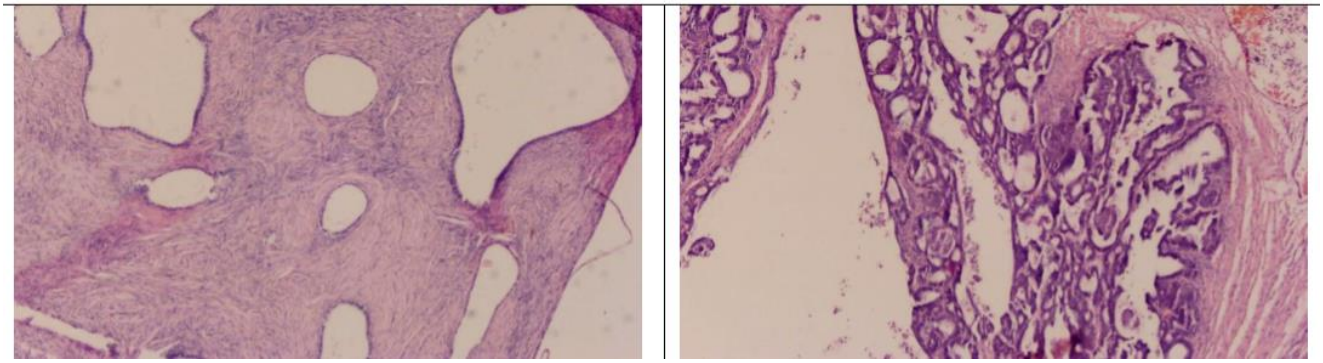


Figure 4. Microscopic appearance of ovarian adenocarcinoma.

Tumor tissue of the ovary consists of poorly defined, confluent, deformed, small or enlarged, papillary, multi-row glandular structures formed by large atypical cells with mucoid cytoplasm, polymorphic, hyperchromic nuclei with many mitotic figures. Between the tumor glands there are foci of atypical squamous epithelium with

keratinization. The stroma is expressed unevenly, with lymphoid infiltration, sclerosis, extensive necrosis, hemorrhages.

Conclusion. Klippel-Trenaunay syndrome presents significant difficulties not only for diagnosis, but also for treatment, is a type of systemic dysplasia and deserves attention as one of the many hereditary diseases that affect various tissues of the body, including the internal organs of patients. With the combined occurrence of this syndrome with oncopathology, the course of both diseases can be aggravated, which is reflected in the difficulty of treating both oncopathology and the Klippel-Trenaunay syndrome. This syndrome is a rare hereditary pathology, which is quite severe in relation to the quality of human life. Therefore, knowledge of the clinical manifestations, differential diagnosis, and the choice of management tactics for this severe group of patients are of particular importance for physicians at all levels of medical care.

Ethics approval and consent to participate - All patients gave written informed consent to participate in the study.

Consent for publication - The study is valid, and recognition by the organization is not required. The author agrees to open publication.

Availability of data and material – Available.

Competing interests – No.

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Conflict of interests - The authors declare that there is no conflict of interest.

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