**Results** Biopsy of the lesions concluded to be a vulvar lymphangioma associated with squamous cell carcinoma of the right labia. The tumour was classified as FIGO stage IA and the patient had a total vulvectomy with bilateral inguinal lymphadenectomy. The evolution was without recurrence of either tumour or lymphangiomatous disease after 6 months.

**Conclusion** Vulvar lymphangioma is a troublesome condition that may prompt a search for an underlying obstructive cause when acquired. It poses a problem of diagnosis and management. The particularity of our case is the presence of an associated micro invasive squamous cell carcinoma of the vulva.

---

**Introduction/Background** Intestinal-type carcinoma represents a rare vaginal primary tumor. The most supported hypothesis of the histological genesis is that the tumor arises from congenital so-called ‘cloacal remnants’. It generally appears as a polypoid vaginal mass in women of 50 years on average. Only ten cases are reported. Usually, the described lesion was small and women underwent excisional surgery and/or radiotherapy.

**Methodology** Case report and literature review.

**Results** We describe a case of a 58-year-old woman who came to our attention with a 10 cm uterine mass involving the uterine cervix and corpus, extending to the upper half of the vagina and infiltrating the anterior wall of the rectum, up to 1 cm from the anal verge, as well as part of the levator ani muscle, close to the left pelvic wall. She previously underwent a uterine biopsy showing an endometrioid adenocarcinoma of unknown origin. The CT scan and the PET/CT uptake confirmed the presence of locally advanced disease, and the patient underwent 5 courses of neoadjuvant chemotherapy with carboplatin and paclitaxel. After the fifth cycle she developed a perineal abscess arising from the utero-rectal component of the tumor, shrived by the chemotherapy. After surgical drainage and antibiotics the abscess resolved. Thereafter, considering the partial response to chemotherapy, she underwent a posterior pelvic exenteration with definitive colostomy and bilateral V-Y flaps reconstruction. The final pathology showed a mucinous intestinal-type carcinoma of the vagina with microscopic infiltration of the left levator ani. She was proposed for adjuvant radiotherapy.

**Conclusion** Since vaginal intestinal-type neoplasms are morphologically similar to their counterpart of the intestinal tract, the issue is how to differentiate between primary vaginal cancer and metastases. Although very rare, awareness is important to avoid misdiagnosis and accurate pathological analysis is essential, as like as the personalized oncological approach to every single case.