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Pleomorphic Vulvar Leiomyoma with Local Invasive Behavior

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A B S T R A C T

A case of pleomorphic leiomyoma in Bartholin gland's area in a 26-year-old woman is reported. After diagnostic treatment, primary excision was done. A large, solid tumor 10 7.5 cm was extirpated. The tumor showed locally invasive behavior, which suggested a malignant tumor of Bartholin gland, because of it's localization and outlook. Pathohistological examination and immunohistochemical reactions proved that it was a mesenchymal tumor of smooth muscle origin with marked polymorphism, without mitosis, with a myxoid stroma and with biological aggressivity, and the possibility of local recurrence. Thus, a second more radical surgical procedure, was performed. In the excised tissue, no residual tumor was found and all lymphnodes were negative.

Introduction

Vulvar leiomyoma is very rare. Especially rare is pleomorphic or bizarre leiomyoma in the vulvar area. Cases of bizarre or pleomorphic leiomyoma of some other localization (kidney, stomach, prostate) were still reported, but bizarre vaginal leiomyoma[1] and vulvar (Bartholin's gland area) leiomyoma[2] were reported in only few cases. This, usually benign tumor, is very hard to differ from leiomyosarcoma. The main criterion to find a difference between them is the mitotic activity, which is less than 3 per 10 high power field (HPF) in atypical leiomyoma. This tumor has not only bizarre outlook, but can have unpredictable behavior[3].

The symptoms are often scanty (dyspareunia and painful swelling in the area of labia mayor and introitus). The therapy is a surgical procedure. Its degree depends on histological and clinical estimate of tumor behavior.

Case Report

A 26-year-old woman, gravida 2, para 1, was referred to the Department of Gynecology and Obstetric at the University Hospital »Osijek« for the evaluation of

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suspicious paravaginal tumor in distal vaginal third and right vulvar half area. The patient has recognized the tumor in distal vaginal third and right vulvar half area six months earlier. In the last few weeks the swelling started to be more painful, and caused dyspareunia, because of the narrow vaginal introitus. The patient had no history of any earlier diseases. Clinically she had a hard tumor mass, painful on palpation, located in distal vaginal third and below the right labia mayor. Other gynecological and ultrasound examination of the uterus and ovaries, PAP smear and blood tests revealed normal findings. Surgical procedure was done. By vaginal and lateral perineal incision we got to the tumor, which was partly enucleated. In its cranial part the tumor tissue infiltrated the surrounding tissue, and a sharp excision of the tumor was done. Large solid tumor with an uneven surface, 10 7.5 cm big and two pieces of the tissue, which content parts of the tumor were removed. The cross sectional tumor tissue is soft, solid yellow colored and partly necrotic. During the excision and enucleation we find out local invasive tumor behavior, which, with concerning on localization and macroscopic look suggested that it was a malign tumor of Bartholin gland. By histological examination we found out that the tumor was solid, made of bundles mostly with spindle cells. Spindle cells have round or oval nuclei with big, prominent, eosinophilic nucleoli. Nuclei partly show hyperchromasy in places where there are cells with giant nuclei. Cytoplasm are eosinophilic elongated, with small hell vacuole. In this area cells have two or more nuclei, with earlier mentioned polymorphism (Figure 1).

Between the cell bundles there is hyaline degeneration, and areas with plenty of myxoid Alcian positive intercellular substance (Figure 2).

Vascularization is in shape of many blood vessels with a thin wall. Mitosis were not found, although the tumor was examined by three independent pathologists from three different countries. The tumor has no capsule, on the margins growth looks infiltrative (Figure 3).

By immunohistochemical procedures carried out in two independent institution for gynecologic pathology proved positive reaction on vimentin, smooth muscle actin and desmin, which refer to a smooth muscle origin of the tumor. Citokeratin MNF 116, EMA, S–100, CD–34 and CD–117 are negative.

Based on established immunofenotype of tumor cells and histological and macroscopic examine, the conclusion is that it is a mesenchymal smooth muscle tumor, with marked polymorphism, without mitosis, with myxoid changes in the stroma and with infiltrative margins.

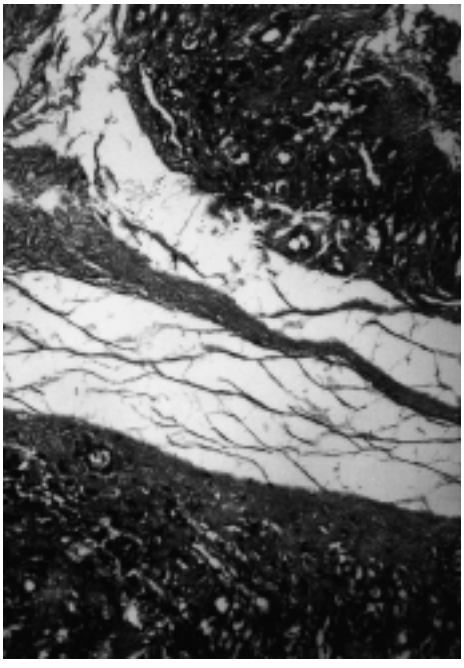

Fig. 1. Pleomorphic leiomyoma – polymorphism of cells and nuclei, HE 400 .

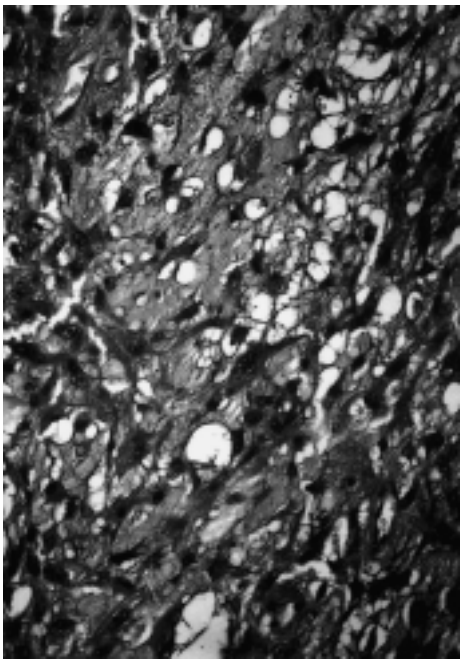

Fig. 2. Pleomorphic leiomyoma – tumors area with myxoid changes, HE 100 .

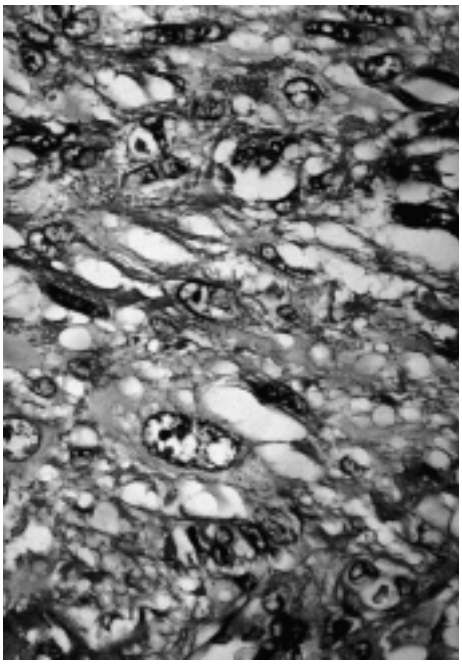

Fig. 3. Pleomorph tumor cells near blood vessel, outside of the tumor, point out at invasive growth of tumor, HE 100 .

According to the pathohistologists opinions, the tumor's size, his outlook and cell pleomorphism, suggest that the tumor is biologically aggressive and local recurrence or eventually metastasis can be expected. The pathohistologists suggestion was to perform a second surgically procedure seven weeks after enucleation and excision. This procedure was a resection of the tumor site, ipsilateral inguinal and femoral lymphadenectomy, besides the parailiacal lymphadenectomy on the right side, by extraperitoneal access. In the excised material from the tumor site area a tubuloalveolar mucous glandula – glandula Bartholin was found and histologically it was normal. In the inguinal, femoral and parailiacal lymphnodes and in the material from re-excised site of the tumor by histological examination the tumor tissue was not found. Fifteen months later, the patient had no local or regional disease recurrence. The patient will be followed-up in gynecologic-oncologic out patient department.

Discussion

There is still one case of bizarre leiomyoma in Bartholin glands area reported in literature[2], but this, usually benign tumor, sometimes is very difficult to differ from leiomyosarcoma of the same area[3].

The appearance and quick growth of leiomyoma of Bartholin gland's like it was documented and in literature earlier described, may be connected with a hormone replacement therapy[4]. Our patient did not receive any hormone replacement therapy.

It is very often a simultaneous development of leiomyoma of the uterine corpus and leiomyoma from the labium ma-

ius[5]. According to clinical examination and by ultrasound the same uterus tumor was excluded.

Of three main determinants of the malign neoplasm of mesenchymal origin (>5 cm, > 5 mitosis/10 HPF and infiltrative margins)[6], the tumor in our case was 10 7.5 cm with infiltrating margins, but mitosis were not found.

There are three main histological criteria for the diagnosis of leiomyosarcoma: coagulation necrosis of the tumor cells, significant, usually diffuse, nuclear atypia and frequent mitotic figures (MF), often atypical.

However, disagreement about the importance of particular criteria on biological behavior of the tumor exist. Now, it is clear that one of the important criteria beside the frequent mitotic figures and cellular atypia is the coagulation necrosis of the tumor cells, and perhaps the most important particular prognostic factor for biological behavior of leiomyosarcoma. In practice, there is a group of smooth muscle tumors that could not be put into true benign nor into true malignant tumors. They are called smooth muscle tumors of uncertain malignancy. The recognition criteria for this group of tumors defined by J. A. Benda and R. Zaino are:

- cytological atypia, 2–5 MF/10 HPF;
- hypercellularity, without atypia, 5–10 MF/10HPF;
- normocellularity, without atypia, 10–15 MF/HPF;
- infiltrative margins, 5–10 MF/10 HPF;
- atypical mitotic figures and/or coagulation necrosis;
- cells of epitheloid type, 2–5 MF/10 HPF;
- symplastic tumor, 2–5 MF/10 HPF;
- vascular invasion, 2–5 MF/10 HPF;
- intravenous leiomyomatosis, 2–5 MF/10 HPF;
- parasitic leiomyoma, 5–9 MF/10 HPF[3].

The presented tumor content cytological atypia, hypercellularity, giant cells which are characteristic for symplastic tumors and infiltrative margins, but do not content one of the most important element – frequent mitotic figures. This is the main reason why we defined this tumor as pleomorphic (atypical) leiomyoma, and not as smooth-muscle tumor of uncertain malignant potential, although its biological behavior is more likely to be an uncertain malignant potential tumor.

Also by WHO definition, bizarre leiomyoma are the tumors that content giant cells, with pleomorphic nuclei with little or without mitotic activity[7].

Myxoid changes in vulvar leiomyomas are common and also related to its aggressive behavior and recurrence[8]. The presented tumor shows in hyaline stroma and in some cross-sections myxoid stroma.

Because of all these elements, this tumor is, according to pathologists opinion, estimated as a biologically aggressive tumor, so we can expect, because of tumors stroma myxoidity, infiltrative margins, cellular polymorphism and tumor size, local recurrence or even metastasis. This is why we did more radical surgical procedure.

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PLEOMORFNI LEOMIOM VULVE S LOKALNO INVAZIVNIM PONAŠANJEM

S A Ž E T A K

Prikaz pleomorfnog leomioma u području Bartolinijeve žlijezde u dvadesetšestogodišnje žene. Nakon dijagnostičkog postupka učinjena je primarna ekscizija. Odstranjen je veliki solidni tumor, 10 7,5 cm. Tumor je pokazivao lokalno invazivno ponašanje, koje je zbog lokalizacije i izgleda tumora sugeriralo da se radi o malignom tumoru Bartolinijeve žlijezde. Na osnovi patohistoloških i imunohistokemijskih pretraga zaključeno je da se radi o mezenhimalnom tumoru porijeklom od glatkog mišićja, s izraženim polimorfizmom, bez mitoza, s miksoidnom stromom i biološkom agresivnošću i mogućnošću nastanka lokalnog recidiva. Zbog toga je učinjen još jedan kirurški zahvat radikalniji od prethodnog. U ekscidiranom tkivu nije nađen rezidualni tumor i svi limfni čvorovi su bili negativni.