Disseminated Peritoneal Leiomyomatosis

Case Report

Mozhdeh Momtahan[1], Maryam Nemati[1], Akbar Safaei[2]

Abstract

Leiomyomatosis peritonealis disseminata is a very rare condition characterized by the development of multiple smooth muscle-like nodules in the peritoneal cavity. It is associated with increased serum levels of gonadal steroids. The present report describes a 29-year-old patient underwent transabdominal hysterectomy and Bilateral Salpingo oophorectomy six years ago because of leiomyomatosis peritonealis disseminata. After six years she referred to us again because of retroperitoneal fibroma, another rare entity, during hormone replacement therapy inspite of lack of uterus and previous castration.

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Keywords ● Leiomyomatosis ● oral contraceptive ● uterine leiomyoma ● mesenchymal stem cells ● metaplasia

Introduction

Leiomyomatosis peritonealis disseminata (LPD) is a benign condition, and a very rare disease. It is suspected that this disease originates from a metaplasia of submesothelial multipotent mesenchymal cells.[1,2] Since female gonadal steroids play an important role in the pathogenesis of LPD, it is generally associated with high levels of exogenous and endogenous female gonadal steroids.[3-6] The definite treatment of LPD is transabdominal hysterectomy and bilateral salpingo oophorectomy (TAH-BSO).[7] However we here describe a case that in spite of TAH-BSO and removal of all peritoneal myomatous nodules, developed retroperitoneal fibroma six years later.

Case Description

The patient was a 29-year-old woman who complained of the presence of a mass in her lower abdomen, and a right flank pain for the preceding six months. The pain increased gradually, and the patient referred to hospital. At clinical examination, the patient didn't present abdominal distention. She, however, had a diffuse pain, which was most intensely observed in the right lower quadrant. Ultrasonography revealed a large (79×45 mm) solid oval shape and well-defined hypoechoic mass in the right adnexal site, which most likely was a residue or recurrence of a previously resected pelvis mass. Six years earlier, due to diffuse and progressive abdominal pain she had undergone abdominal ultrasonography, which revealed a semi-solid mass (54×27 mm) in the left side of adnexa attached to uterus. She was then subjected to laparotomy to remove the mass. The laparotomy revealed a very fragile, largely vascular and multi nodular solid mass, which had originated in posterior part of uterus and extended to peritoneum and retroperitonem. The macroscopic presentation of the mass mimicked a disseminated malignancy. TAH-BSO were performed because of intractable bleeding following

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the resection of a retroperitoneal mass. The microscopic pathology findings confirmed the mass as leiomyoma. For more than 5.5 years after the surgery, the patient was doing well with no recurrence of the tumor. However, flank pain and mass sensation started and persisted during hormone replacement therapy since six months ago. She underwent the second operation six months ago, and a retroperitoneal solid mass (6×8 mm) with an irregular border and a pseudo-capsule was found just adjacent to the external iliac artery. Histopathogical examination of the pelvic mass exhibited interlacing bundles of smooth muscle cells without cytological atypia, and a few mitoses (figure 1). Immunohistochemical evaluation was strongly positive for the smooth muscle antigen (figure 2), progesterone receptors (figure 3), and estrogen receptors (figure 4), but was negative for cytokeratin. Histopathogical and Immunohistochemical findings were in favor of uterine leiomyoma. Considering the patient's history, the mass was suggested to be a retroperitoneal fibroma, a remenant of previous disseminated peritoneal leiomyomatosis. The patient was, therefore, scheduled for a long-term follow-up.

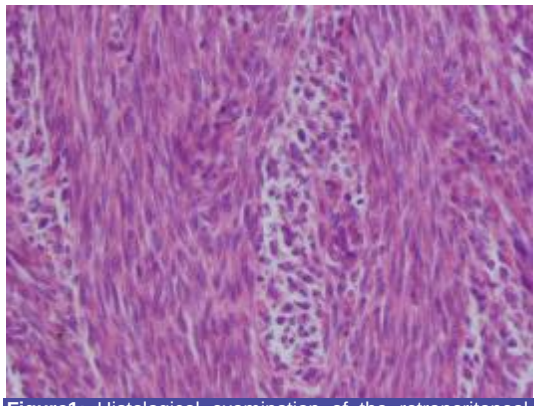


Figure1: Histological examination of the retroperitoneal mass show bundles of spindle cells with eosinophilic cytoplasm (H&E).

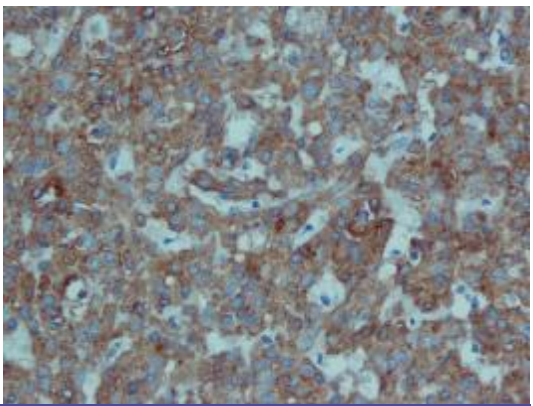


Figure2: The immunohistochemical evaluation is strongly positive for smooth muscle actin (cytoplasmic staining).

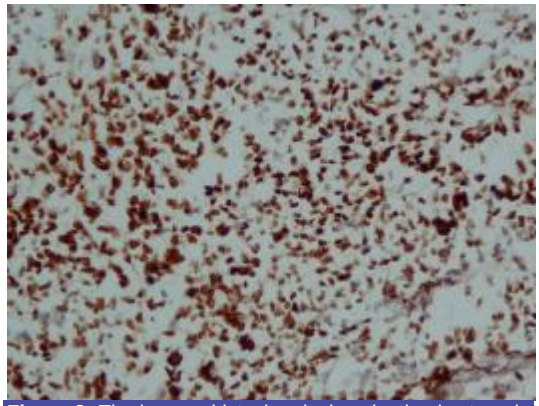
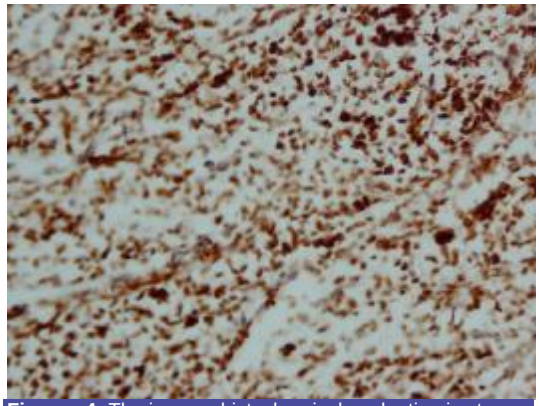


Figure 3: The immunohistochemical evaluation is strongly positive for progesterone receptors(nuclear staining)



Figures 4: The immunohistochemical evaluation is strongly positive for estrogen receptors(nuclear staining).

Discussion

LPD is a very rare and benign disease, of which less than 140 cases have been reported.[8] It is characterized by the presence of multiple smooth muscle nodules in the peritoneal surface. The disease etiology still remains unknown. Some authors believe that female gonadal steroids play an important and primary role in the pathogenesis of leiomyomatosis peritonealis. Similar to a number of situations such as pregnancy, extended exposure to oral contraceptives and/or combined hormonal replacement therapy, oestrogen-secreting ovarian fibrothecoma, the disease is generally associated with high levels of exogenous and endogenous female gonadal steroids.[3-5] Since, the disease is seen in post menopausal women and in males, the possible causes of it could be divided into hormonal, subperitoneal mesenchymal stem cells metaplasia, genetic, or iatrogenic after morcellation of myoma during laparoscopic surgery.[2,8] Surgeans and pathologisis face a challenge when dealing with LPD because its macroscopic appearances

resembles peritoneal carcinomatosis. While this condition is not common, it has to be taken into account in dealing with a patient with abdominal masses, and especially after a previous myomectomy or hysterectomy.[1,2,4,5] The diagnosis of LPD is made on biopsy specimen, which generally demonstrates a benign smooth muscle abnormal growth originating from the multicentric metaplasia of the peritoneal surface.[8-11] LPD needs to be distinguished from leiomyosarcoma.[4,5,8] Steroid hormone receptors have been detected in the proliferating cells of LPD.[1,3,5] So the reduction of estrogen exposure results in regression of LPD.[1,3,9] Therefore surgical casteration or gonadotropin releasing hormone agonist seems the appropriate treatment of such conditions.[3,12]

Conclusion

Leiomyomas should be considered in the differential diagnoses of intraperitoneal or retropevitoneal masses distinct from the uterus.

Conflict of Interest: None declared

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