Leiomyomatosis peritonealis disseminata in a postmenopausal woman

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Leiomyomatosis peritonealis disseminata is an unusual and rare entity; 47 cases have been reported to date. Of those cases, only three were described in postmenopausal women (1–3). Two cases were described in previously hysterectomized women (2, 4).

Most of the cases were accompanied by estrogen increase; due to estrogen-secreting granulosa cell neoplasm, pregnancy or oral contraceptive use (5). Usually, most of the cases were found incidentally, during laparotomies performed for other reasons. Here we present an unusual case of LPD in an 82 year old obese menopausal woman with adnexa and uterus completely removed 40 years ago, with no evidence of endometriosis at that time.

Clinical summary

An extremely obese, Caucasian female, gravida 2, para 2, presented to Clinical Hospital Split because of diffuse pain in the abdomen. Previous medical history revealed uterine leiomyomata with consecutive hysterectomy and bilateral salpingooophorectomy forty years ago. No endometriosis was identified during the laparotomy. She had never used oral contraceptives.

Two nodular masses were identified in the pelvic cavity, by means of ultrasound and CAT scan, measuring 10 cm and 3.5 cm in greatest diameter, respectively. Preoperative routine diagnostic studies were in normal range except increased serum level of glucose (8.62 mM/L).

Surgery was performed, and two previously described nodular masses were found in the pelvic region. In addition, multiple nodular masses were found on the surface of the large and small bowel, urinary bladder and right broad ligament, ranging from 0.3 cm to 2 cm in the greatest diameter. All nodular lesions were entirely excised.

Abbreviation:
LPD: leiomyomatosis peritonealis disseminata.

Pathohistological findings

Several tissue blocks from each of the nodular lesions were fixed in formalin and routinely processed for light-microscopy examination. Histologic findings revealed well circumscribed neoplastic nodules, composed mainly of spindle cells. The cells were arranged in whorls and had an abundant eosinophilic fibrillar cytoplasm. Elongated nuclei were observed that contained small distinct nucleoli, all characteristic of the smooth muscle cells. Mitotic figures were absent, no overt atypia was seen, and the cells were not hyperchromatic. Neither necrosis nor hemorrhage were present in any of those nodules.

Neoplastic cells were strongly positive for smooth muscle actin and vimentin, and weakly positive for desmin (DAKO EPOS) (not shown).

Ultrastructural findings

Ultrastructural examination was carried out on the formalin-fixed tissue that was postfixed in OsO_4, dehydrated in series of acetone and embedded in Durcupan. Ultra thin sections were stained with uranyl acetate and lead citrate and analyzed by electron microscope (Opton Em 9A). It was evident that the majority of the cells resemble mature smooth muscle cells with slightly irregular, elongated nuclei (see Fig. 1).

Discussion

LPD is a benign disorder that presents as a multifocal proliferation of smooth muscle like cells. It is hypothesized that multipotent subcoelomic mesenchymal cells proliferate in response to female estrogens, differentiating into myoblasts, myofibroblasts, fibroblasts, and even decidua-like cells (6). In our patient, the majority of the cells resemble mature smooth muscle cells arranged in whorls and bundles (Fig. 1).

Although most of the nodules of LPD are small, measuring a few millimeters in diameter, neoplasms up to 10 cm were described (1, 3, 4). In this study we found two nodules of 10 and 3.5 cm in diameter and multiple nodular masses ranging in diameter from 0.3 to 2 cm. Serosa of the large and small bowel, and pelvic region (where the most of the nodular masses were located), are common sites of involvement. Involvement of two other sites (urinary bladder surface and adnexal region) has also been described (4).

Of the previously reported 47 cases, 23 patients were pregnant at the time of diagnosis, 17 were taking oral contraceptives for many years, one patient had a granulosa-cell tumor, one was male, and the five remaining did not have any history of hormonal increases during their lifetime. Therefore, it is evident that LPD occurs in association with hyperestrogenism.

However, there are three cases of LPD reported in postmenopausal women of 51 (1), 55 (3) and 63 (2) years old. Two cases were in previously hysterectomized women (2, 4). The increasing number of postmenopausal LPD cases, with one reported in a male, may suggest that other factors, besides the hormonal, can play a role in the development of this rare disease.

Our patient was not only postmenopausal, but she also had undergone hysterectomy with bilateral salpingooophorectomy forty years ago, and had never used hormonal contraceptives. There were no endometriotic inclusions in our specimens which would support the theory of metaplasia of the coelomic-derived mesothelium. Therefore, we suggest that the other possible causes should be brought to attention in the future studies. One possibility is peripheral conversion of androgens (she was extremely obese). However, clinical features and laboratory investigation revealed neither hyperestrinism nor hyperandrogenism in our patient.

Fig. 1. (EM X 6000) Smooth muscle-derived tumor cell with slightly irregular elongated nuclei. Note cytoplasm rich in rough endoplasmatic reticulum with bundles of parallel myofilaments. Well developed basal lamina is seen on the outer aspect of the cell membranes.

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