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GI IMAGE

Disseminated Peritoneal Leiomyomatosis

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A 53-year-old premenopausal woman was referred to our unit with a two-year history of progressive abdominal pain. The patient was nulliparous and had only benefited from umbilical hernioplasty 10 years ago. Physical examination found moderate pain of the abdomen with abdominal soft distension. No palpable mass was detected. Laboratory tests were normal. Abdomino-pelvic computed tomography (CT) and magnetic resonance imaging revealed massive infiltration of the lower parts of the abdomen by multiple enhancing solid and gelatinous lesions, covering all structures from the pelvis to the umbilic area without invading or compressing the intestine (Fig. 1), suggesting a benign form of pseudomyxoma peritonei such as peritoneal adenomucinosis. Thoracic CT found a similar 3-cm long cyst in the upper para-tracheal region. Colonoscopy was normal. Laparoscopic exploration was finally conducted and found, instead of classical puddles of mucin, multiple soft nodules of various sizes, massively infiltrating the great omentum, pelvis, pericolic gutters and the lower parts of the peritoneum, but preserving the small intestine (Fig. 2). Histopathology identified highly cellular interlacing bundles of smooth muscle cells, mimicking a leiomyosarcoma, although there were no nuclear atypia, mitotic figures, or signs of necrosis. Immunochemical analyses confirmed that the lesion was not malignant with a Ki67 ratio of 0%, intensively expressing desmin, caldesmon, estrogen, and progesterone receptors. A diagnosis of disseminated peritoneal leiomyomatosis was established. Because the disease was suspected to be metastatic and no signs of organic compression were noted, we decided to not perform surgical resection and proposed hormonal therapy using aromatase inhibitors. At one-year follow-up, the patient was fit and well, and imaging exams demonstrated stabilization of the disease.

Disseminated peritoneal leiomyomatosis (DPL) is a rare disorder first described by Willson and Peale in 1952,[1] characterized by widespread of myofibroblastic nodules in the peritoneal cavity, mimicking the peritoneal seeding of a malignant tumor. Although DPL has been described in postmenopausal women, the condition usually affects active women and has been associated with pregnancy, uterine leiomyoma, or prolonged oral contraceptive use. The pathogenesis of DPL is unclear but, according to Tavassoli and Norris,[2] DPL results from the transformation under hormonal stimulation of subperitoneal mesenchymal stem cells which exhibits predisposition to metaplasia. This theory is supported by the fact that DPL is associated with other metaplastic lesions such as endometriosis or endosalpingiosis. An iatrogenic cause of DPL has also been suggested, notably when power morcellators are used during laparoscopic surgery of uterine fibroids, as a result of the widespread fibroid pieces in the peritoneal cavity. Extraperitoneal localizations as in our case, however, suggest other possible means of dissemination.

Preoperative diagnosis of DPL is challenging. As the disease is very little known, imaging studies usually shift towards peritoneal carcinoma or pseudomyxoma peritonei. The indolent course of the disease and the absence of an altered general condition usually suggest a borderline disease, but only histo-

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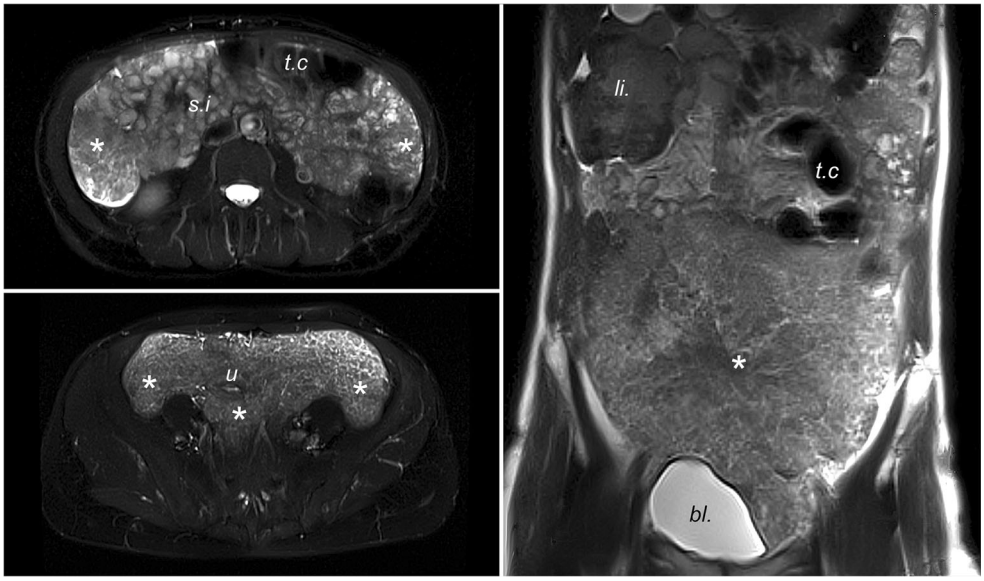

Fig. 1 MRI (T2 axial and coronal views), showing massive infiltration of the lower parts of the abdomen by heterogeneous lesions (asterisks); t.c, transverse colon; s.i, small intestine; u, uterus; li., liver; bl., bladder

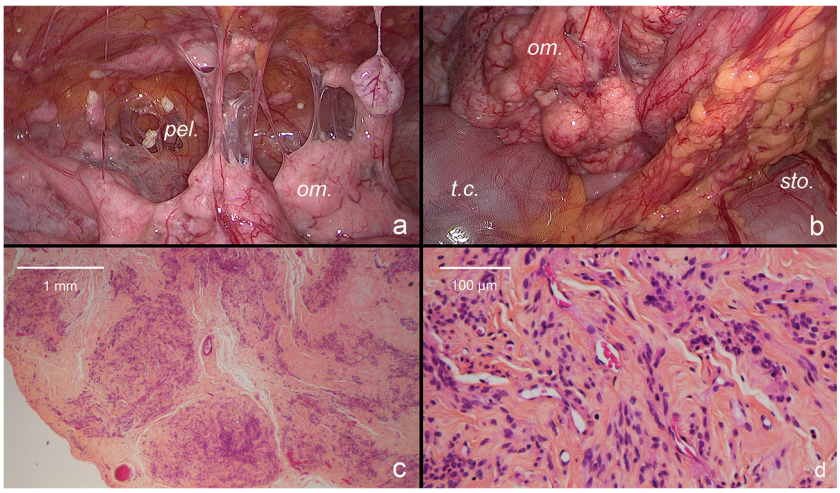

Fig. 2 Macroscopic (laparoscopic) views of the abdomen (**a**, **b**); HES × 2.5 showing groups of cells disposed on a fibrohyaline background (**c**); HES × 20 showing fusiform cells without nuclear atypia (**d**). om., omentum; pel., pelvis; t.c, transverse colon; sto., stomach

logical and immunohistochemical studies can confirm DPL, showing smooth muscle cells without nuclear atypia, strongly expressing estrogen and progesterone receptors. The evaluation of mitotic activity using the Ki67 index usually confirms whether the lesion is pauci proliferative or not, conversely to other malignant disorders.

The optimal management of DPL remains unclear. Because rare cases of malignant transformation have been described, some authors recommend up-front surgery with peritoneal debulking and hysterectomy/ovariectomy. We do not support this opinion because: (i) neoplastic transformation of DPL is uncertain, supported by few reports, which do not prove an actual connection between DPL and sarcoma; (ii) according to the hormonal theory, resection surgery would be ineffective in the cause of DPL and could not prevent recurrence; (iii) peritoneal debulking is an aggressive strategy, particularly if lymphadenectomies are discussed for treatment of other possible means of metastatic dissemination; (iv) hormonal therapy using aromatase inhibitors or gonadotropin-releasing hormone agonists has proven to be effective, acting as a local and systemic controller.[3] Extensive surgery should thus only be considered in cases of organic compression. Up-front resection of DPL should be avoided and preceded by hormonal therapy administrated in a neo-adjuvant setting in order to preoperatively control the disease and limit the extent and morbidity of the surgical resection.

Author Contributions Clément Julien and Stéphane Bourgouin drafted the manuscript. Laurys Boudin and Paul Balandraud critically revised the manuscript.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Willson JR, Peale AR. Multiple peritoneal leiomyomas associated with a granulosa-cell tumor of the ovary. Am J Obstet Gynecol 1952; 64: 204–8.
2. Tavassoli FA, Norris HJ. Peritoneal leiomyomatosis (leiomyomatosis peritonealis disseminata): a clinicopathologic study of 20 cases with ultrastructural observations. Int J Gynecol Pathol 1982; 1: 59–74
3. Hales HA, Peterson CM, Jones KP, Quinn JD. Leiomyomatosis peritonealis disseminata treated with a gonadotropin-releasing hormone agonist: a case report. Am J Obstet Gynecol 1992; 167: 515–6.