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Benign multicystic peritoneal mesothelioma: case reports and review of the literature

Mesotelioma peritoneale multicistico benigno: casi clinici e revisione della letteratura

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Summary

Benign multicystic peritoneal mesothelioma is a very rare benign cystic tumor; at present approximately 130 cases have been reported. It more frequently occurs in females (87%), expecially in the pelvic area of the cul-de-sac, uterus and rectum; the pathogenesis has remained controversial. Abdominal pain is the most common symptom (75%). Ultrasound, Computerized Tomography (CT) and magnetic resonance imaging are helpful, but the diagnosis is confirmed by histopathological and immunohistochemical examination. Benign multicystic peritoneal mesothelioma shows high recurrence after surgical resection, but it does not present a tendency to transform into malignancy. Surgery is the only effective treatment, and peritonectomy is recommended. A prolonged systematic follow-up of these patients, perhaps for all life, is

Riassunto

Il mesotelioma peritoneale multicistico benigno è una patologia molto rara, di cui in letteratura sono attualmente riportati circa 130 casi. È più comune nelle donne (87%), specialmente nell'area pelvica del cul-de-sac, utero e retto; la patogenesi rimane controversa. L'esordio più comune della patologia è il dolore addominale (75% dei casi). La diagnosi viene generalmente sospettata tramite un'ecografia, una Tomografia Computerizzata (TC) o una Risonanza Magnetica (RM), mentre la risposta definitiva si ha tramite l'esame istologico e immunoistochimico. Si tratta di lesioni generalmente benigne, con alta incidenza di recidiva locale e scarsa tendenza alla disseminazione a distanza. L'unica terapia efficace è la chirurgia, che dovrebbe essere il più possibile radicale e dovrebbe comprendere anche la rimozione del peritoneo. Rimane indicato perciò un

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required. Here we report two cases that underwent surgery for benign multicystic peritoneal mesothelioma. In the first case the lesion was a multicystic fluid mass of the large intestine, while in the second one a smaller peritoneal mass was reported. In both cases the pathological result was a benign multicystic peritoneal mesothelioma. After surgery, we decided to continue with a prolonged systematic follow-up. Eur. J. Oncol., 15 (2), 119-123, 2010

Key words: **benign multicystic peritoneal mesothelioma, peritoneal tumor**

costante e attento follow-up, verosimilmente per tutta la durata della vita dei pazienti. Riportiamo due casi di mesotelioma peritoneale multicistico benigno. Nel primo caso era presente una lesione multicistica a contenuto fluido a partenza dal piccolo intestino, mentre nel secondo era presente una piccola lesione peritoneale. Entrambi sono stati sottoposti ad intervento chirurgico di asportazione di lesioni cistiche peritoneali: si trattava di due casi di mesotelioma peritoneale multicistico benigno. Dopo l'intervento chirurgico, si è deciso di seguire le pazienti con un prolungato e sistematico follow-up. Eur. J. Oncol., 15 (1), 119-123, 2010

Parole chiave: **mesotelioma peritoneale multicistico benigno, neoplasie peritoneali**

Introduction

Benign multicystic peritoneal mesothelioma (BMPM) is a rare tumor. It was firstly described in 1979 by Mennemeyer and Smith (1); at present approximately 130 cases have been reported (2), expecially isolated case reports. This lesion more frequently occurs in women during their reproductive years and is associated with a history of previous abdominal surgery (60%), endometriosis (36%) or pelvic inflammatory disease (28%) (3-8); however, cases concerning men or children, as well as rare extra-abdominal cases are reported (9). Moreover cases with history of a previous cesarian section are reported (10).

BMPM is a rare benign cystic tumor, with high recurrence after surgical resection, but it does not present a tendency to transform into malignancy. Surgery is the only effective treatment for BMPM (11).

Case 1

We report a case of a childless 72-year-old-woman, with a history of Hashimoto disease and breast cancer (1995, ductal carcinoma, pT1N0M0) treated with surgery and radiotherapy. In June 2008 she presented abdominal pain. Ultrasonography (US) showed a large, intraperitoneal, multicystic fluid mass (8 cm of diameter). Subsequent Computerized Tomography (CT) scan (fig. 1) confirmed a multicystic fluid mass of the large intestine, in first hypotesis an appendiceal mucocele. In June 2008 the patient underwent open abdominal surgical exploration; complete resection of the lesion was performed. The pathological result was a benign multicystic peritoneal mesothelioma; cytologic examination of the peritoneal fluid was negative for neoplastic disease.

Therefore the patient was admitted to our Clinical Oncology Unit. Because of high recurrence after surgical resection and the very rare tendency to transform into malignancy, we decided to continue with a prolonged systematic follow-up with US and CT, without any specific oncological treatment. After more than 1 year, the patient is actually free of recurrence.

Pathology

Microscopic examination shows one or more variously sized, round or irregularly shaped cystic spaces lined by a single layer of flattened or cuboidal mesothelial cells (fig. 2). Focal squamous metaplasia is occasionally observed. The tissue that separate cystic spaces is often edematous and is infiltrate by

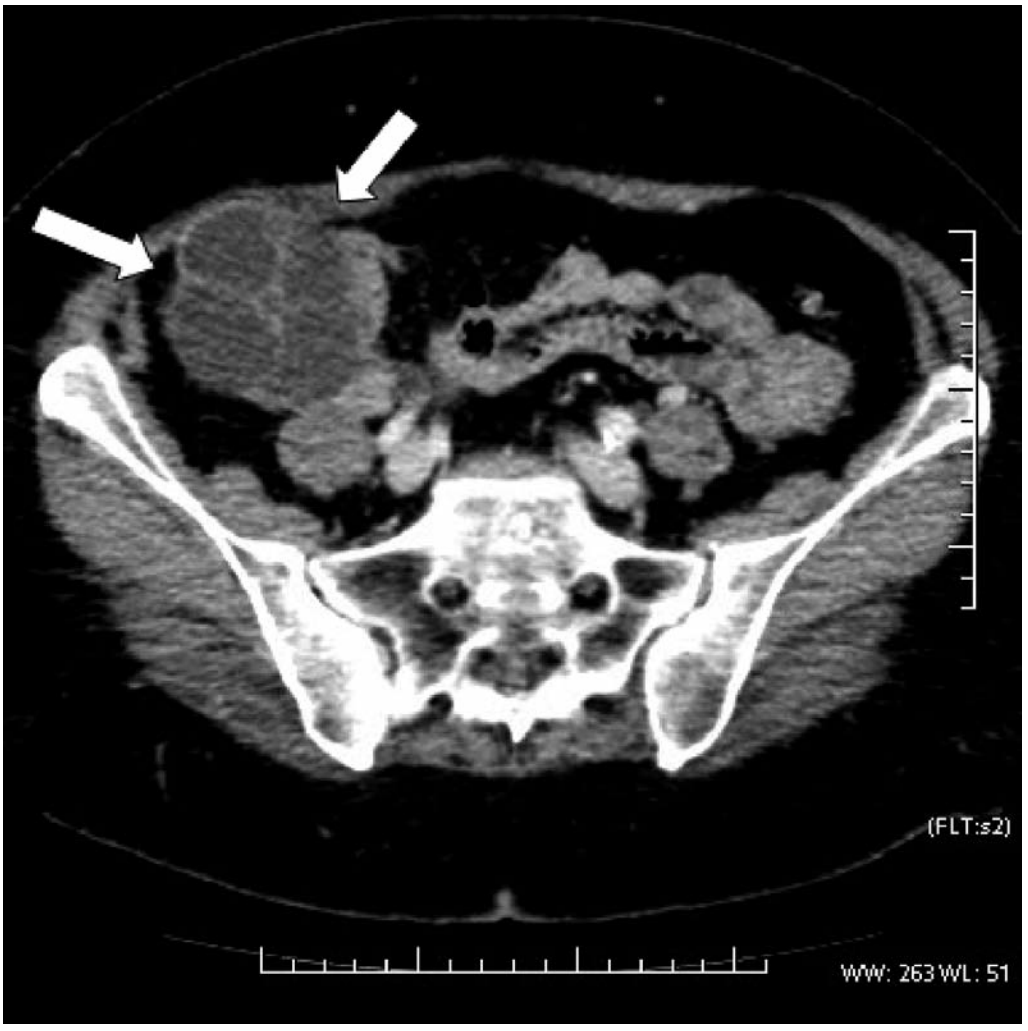

Fig. 1. Case 1: CT scan showing a multicystic fluid mass of the large intestine, apparently divisible from the blind intestine

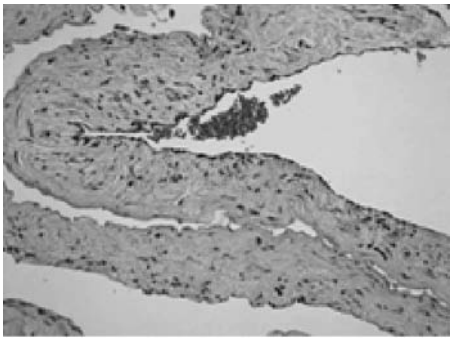

Fig. 2. Case 1: wall lined by flat mesothelial epithelium (H&E 10x)

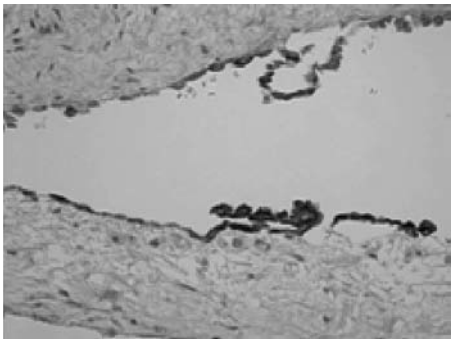

Fig. 3. Case 1: CK5/6 expression (20x)

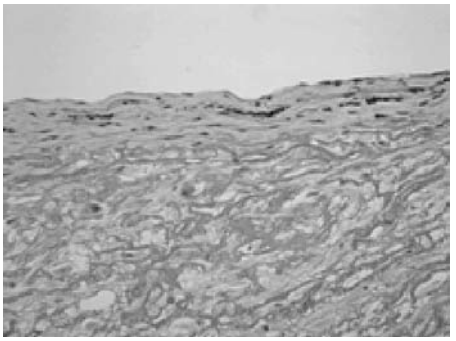

Fig. 4. Case 1: lack of expression of CD31 (20x)

inflammatory cells, fibrin deposits, and sometimes entraps mesothelial cell resembling infiltrating carcinoma (mural nodules). The lining cells express cytockeratin 5/6 (CK5/6, fig. 3). The lack of CD31 expression help to exclude cystic lymphangioma (fig. 4).

Case 2

A childless 26-year-old-woman, presented with intermittent, generalized abdominal pain of two years duration. In April 2009 a smaller peritoneal mass was evident during open abdominal surgical

exploration; complete resection of the lesion was performed. The pathological result was a benign multicystic peritoneal mesothelioma; cytologic examination of the peritoneal fluid was negative for neoplastic disease. Subsequently CT scan does not showed any disease persistence.

Now she is free of symptoms and shows no recurrence. We decide to continue follow-up by US, CT and Cancer Antigen 125 (CA125).

Discussion

BMPM is a rare tumor with annual incidence of 0.15/100,000 population. It more frequently occurs in females (87%), expecially in the pelvic area of the cul-de-sac, uterus and rectum (7, 8); rarely, lesions have been described in the pleura (9), spermatic cord (12), tunica vaginalis (13) and the pericardium (14). It is more common during the third and the fourth decades of life (15); no causative relationship has been found with asbestos exposures, in contrast with malignant mesothelioma (8).

The pathogenesis of BMPM remains controversial. There are two different theories. Some authors have proposed a neoplastic origin based on that this lesion does not metastatise and therefore is not truly malignant, but frequently locally recurs; two cases of malignant transformation have been reported (2, 16). Other authors believe that the lesions are a reactive process related to chronic irritation stimuli, with mesothelial cell entrapment, reactive proliferation and cystic formation (3, 9). The association with previous surgery and endometriosis suggests that the lesion is a reactive process. BMPM was also described in association with Familial Mediterranean Fever (17).

Abdominal pain is the most common symptom (75%); other manifestations may include a mass, an acute abdomen, urinary and/or bowel symptoms, dyspareunia, dysmenorrhea, shoulder pain or even hernia (3, 7, 8). Not uncommonly, diagnosis may be an incidental finding (18% of cases) (3).

Most tumors present a diameter of 4-20 cm (2, 15), but they can grow up till to 30 cm (18).

US, CT and magnetic resonance imaging are helpful, but diagnosis is confirmed by histopathological and immunohistochemical examination (18-20). The differential diagnosis is extremly various and includes any cystic abdominal mass. Benign lesions include cystic endosalpingitis, endometriosis, mullerian cysts involving the retroperitoneum, cystic adenomatoid tumors, pseudomyxoma peritonei and cystic mesonephric duct remnants. Malignancies such as malignant mesotheliomas, serous peritoneal tumors and ovarian clear cell carcinomas may occur (4, 21-23). The most common confusing lesion is a cystic lymphangioma, a men's typical disease restricted to the mesentery, omentum, mesocolon and retroperitoneum but rarely exported in the ovary (3, 24). Pre-operative fine-needle aspiration biopsy of cystic lesions may help the differential diagnosis (25-27). From a clinical point of view, the differential diagnosis of BMPM from cystic tumors of ovaries is most important (20), expecially in ovarian cancer, where biopsy is not recommended. Furtheremore BMPM may be treated through local excision with preservation of the ovaries. On radiological findings cystic ovarian neoplasms occur such as water-density masses in the pelvis; they may be thin-walled and therefore difficult to distinguish from BMPM. Ancillary findings with CT, such as intratumoral mural nodules, ascites, or peritoneal/mesenteric involvement, may suggest ovarian malignancy; however, they are not consistently demonstrated (7).

Surgery is the only effective treatment for BMPM (11); peritonectomy is recommended (28). Adiuvant chemotherapy and radiotherapy are not indicated because these tumors have a prevailing benign character.

Recurrences have been reported at a frequency of 27-75%, with a range of 3 months to 19 years after initial resection (7). Recurrences more frequently occur in women and can be treated with hormonal therapy anti-oestrogens (11, 29) and gonadotrophin-realising analogues (11, 30), hyperthermic intraperitoneal chemotherapy and sclerotherapy (11, 31, 32) with tetracycline (11, 33), with different success. Actually age, sex, site, size, extent and previous recurrence are not helpful in the prediction of the clinical course (1, 3, 8, 15, 34).

Finally a prolonged systematic follow-up of these patients, perhaps for all life, is required (11, 15).

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