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Benign multicystic mesothelioma: a case report of recurrent disease in a young adult with familiarity for renal cell carcinoma

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SUMMARY: **Benign multicystic mesothelioma: a case report of recurrent disease in a young adult with familiarity for renal cell carcinoma.**

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Benign multi-cystic peritoneal mesothelioma (BMCM) is a very rare disease (about 150 cases observed). The aetiology is currently little-known, and the data collected, without having achieved conclusive results, identify two possible causes: neoplastic and reactive inflammatory. This case report refers to a recidivism of BCMC in a patient whose brother, few months before, underwent a left nephrectomy and right renal Radio Frequency Termo Ablation (RFTA) for bilateral papillary renal cell carcinoma. For the recurring trend, the onset in a male young patient without chronic inflammatory diseases evidence, the presence of a first degree relative with a rare carcinoma we supposed a neoplastic aetiology. The available literature suggests that both tumours (BCMC and renal cell carcinoma) are susceptible to oestrogens. This biomolecular mechanism could represent a valid antipathogenic hypothesis.

KEY WORDS: Benign multi-cystic mesotelioma - Surgery - Renal cells kidney tumor - Familiarity - Oncology - Oestrogen.

Introduction

Benign multi-cystic peritoneal mesothelioma (BMCM) is a very rare disease (about 150 cases observed) and the aetiology is still under debate. Plaut et al. described for the first time a case of BCMC in 1928, the mesothelial origin was histologically confirmed by Menemeyer and Smith in 1979. BCMC occurs more frequently in female patients in reproductive age therefore suggesting a hormone-dependence of the same lesion. The male patients' incidence is very low and the majority in paediatric age or over 60 (1). The literature is currently divided but has not yet reached conclusive results, between neoplastic and reactive inflammatory etiologic hypothesis (1-6). The first is suggested by lesion growth rate, high recurrence percentage, low abdominal infections incidence and by high mortality for the extended forms. Some cases of familial BMCM are reported in the literature and several cases of malignant evolution of the lesion which strengthen the onco-genetic hypothesis (1-11). Instead the reactive hypothesis derives from relative higher incidence in subjects with previous peritoneal inflammation outbreaks, endometriosis or uterine leiomyomas and with abdominal surgery history (1, 2, 7, 8).

The diagnostic confirmation is exclusively histological. Many benign or malignant cystic lesions can be put in differential diagnosis with BCMC, like: lymphangiomas, omental cysts, cystic endosalpygiosis, ovarian cystadenoma, ovarian cystadenocarcinoma, cystic teratoma, pseudo-myxoma peritonei, appendicular mucocele, cystadenoma and appendiceal mucinous cystadenocarcinoma, visceral cysts, pancreas mucinous cystic lesions, endometriosis and echinococcal cysts (1, 2, 12-14). Mono-stratified coating presence with cuboidal cells and peculiar

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immunohistochemical aspects (calretinin, cytokeratin 5/6, cytokeratin 7, berep4 focal positivity and negativity for cytokeratin 20, B72.3, CEA monoclonality, and p63) allow an adequate diagnostic certainty (2). Only in 11,7% we can observe a ER positivity and locally for PR despite epidemiological correlation with female sex in reproductive age (2, 6). Studies performed on two cases treated with antioestrogens have shown a cystic lesions regression and a resumption of disease at suspension or after administration of oestrogen and progesterone (2-6). Recently, a case of association between BMCM and Ca19.9 has been observed. Specifically, a high preoperative level and a significative serum marker reduction in the postoperative period have suggested a correlation with BMCM (8). Laparoscopic exploration has in some cases allowed to identify suspicious lesions, not better specified by other instrumental investigations (lesions' translucent appearance, straw yellow content) and in some cases it was possible to perform the laparoscopic excision procedure (8).

Clinical summary

Mr AF (brother 1) was a 48-years-old Caucasian male that underwent, seven years before, surgery for abdominal cystic mass resection, histologically a BCMC, treated without other therapy. A peculiar familiar anamnesis was referred. In fact, three months before the re-hospitalization into our emergency surgery unit, Mr AP (brother 2) was submitted to left nephrectomy and on the right side to neoplastic lesions biopsy, resulted positive for papillary renal cell carcinoma consequently treated with Radio Frequency Termo Ablation (RFTA). In the post-operative period, after first mass resection, Mr AF underwent a periodical Computed Tomography (CT) scan survey. At the last of these CT of the abdomen, compared with a previous one, was observed a significant increase in a cystic lesion (10 x 5.5 x 15 cm) localized in the right iliac fossa in relation to the last ileal loop, cecum and ascending colon. Appearance of further cystic lesion between ileal loops and a millimetric diffuse but unchanged lymphadenopathy. Furthermore, Mr AF refers pain on right lower quadrant. According to literature it was indicated surgery with cytoreductive intent. This is the only therapeutic possibility since hyperthermic peritoneal chemotherapy, proposed from someone, is currently off-label (1, 8-11). Surgical intervention was performed with a starting laparoscopic approach subsequently converted. Medial ileus loop resection with cystic neoformation interposed between the loops and entero-enteric L-L antiperistaltic anastomosis were performed, the cystic antero-cecal lesion was removed en bloc with last ileal loop, cecum and ascending colon, transverse-ileus TL anastomosis with EEA 25 was performed. Finally, the operation ended with the removal of 2 cystic formations in the pelvic excavation not previously found (15-19). In the immediate post-operative period any blood count changes or electrolyte alterations did not show up. On second post-operative day Mr AF showed neither fever nor other abdominal infection signs. The peristalsis resumed on 3rd post-operative day. Patient was re-initiated orally on 4th post-operative day and parenteral nutrition was suspended. In the 5th post-operative day abdominal drainages were removed, and Mr AF was discharged in the 7th post-operative day. After about 15 days, histological examination was received which confirmed BMCM diagnosis and the patient was referred to Oncologist attention. Mr AF has not performed further post-operative therapy. The patient has completely resumed his daily activities and enjoys an excellent performance status.

Discussion

The case of BMCM therefore exposed is added to the already poor world casuistry. It is peculiar because it occurs unusually in a young adult male subject without other remarkable comorbidities and/or inflammatory pathology and with a familiarity for multifocal renal cell tumour.

Recidivism and tumour pathology familiarity presence represent important anamnestic information in the BCMC etiological definition suggesting a probable neoplastic aetiology. Papillary renal cell tumour is associated with several oncogenetic mutations such as those on the VHL/HIF axis (specifically HIF/VEGF signalling pathway seems to play a prooncogenetic role), and those on HGF/Met tyrosine kinase axis whose signal is transmitted through the activation of different Focal Adhesion Kinase

(Ras/Raf/MEK/ERK, and PI3K/Akt). In particular, it should be noted that a chromosome 7 trisomy, in which the gene encoding Met protein is located, is responsible for familial forms of renal cell carcinoma (20). There are literature data that suggest a worse prognosis in case of oestrogen receptors expression by renal tumour cells (21, 22). According to a recent study by Qiang Wang et al. the expression of ER alfa36 represents a fundamental element for the differential diagnosis between benign and malignant renal injury and is correlated with a worse prognosis in case of renal cell carcinoma (8). Furthermore, the increase of the signal by the oestrogen beta receptors would be associated with a greater migration of the neoplastic cells by induction of the 2alpha VEGF/HIF pathway and a worse prognosis (22-25). All this could be correlated with a family estrogenic sensitivity that would justify the BMCM appearance in a young adult male (unusual age). The confirmation of this hypothesis can only be obtained through appropriate family genetic counselling, which our patients have just started.

Conclusions

The BMCM reported case increases the poor world experience on this type of tumour, furthermore its importance is growth by the neoplastic familiarity of multifocal kidney neoplasia. A familiar hypersensitivity to oestrogen could justify the appearance of a case of BMCM in an adult young man (uncommon age). The confirmation of this hypothesis could be obtained only with a specific genetic counselling.

Conflict of interest

Nothing to declare.

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