

A Mature Cyst Teratoma with Malignant Transformation in Squamous Cell Carcinoma Presenting Like a Sigmoid Fistula: A Case Report of a Rare Differential Diagnosis of a Giant Colonic Diverticulum

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ABSTRACT

Mature cystic teratoma is the most common form of ovarian germ cell tumor. Mature cystic teratoma is a benign tumor, mainly diagnosed in young women, with non-specific symptoms. However, the risk of malignant transformation exists (1-2%). Squamous cell carcinoma is the most frequently encountered histological form. Giant colonic diverticulum is a rare complication of diverticular disease, characterized by a diverticulum larger than 4 cm which can sometimes be confused with other digestive pathologies (appendicitis, small bowel perforation ...). We report a case of an ovarian squamous cell carcinoma arising from a mature cystic teratoma, presenting like a sigmoid fistula, with an initial doubt with the diagnosis of a giant colonic diverticulum.

A 68-year-old woman was admitted for chronic hypogastric abdominal pain with a doubt concerning the diagnosis between a mature cystic teratoma and a giant colonic diverticulum. A computed tomography scan was performed revealing a perisigmoid cyst with hydro-aeric level in favor of a giant colonic diverticulum. The pelvic MRI rather suggested a left ovarian cyst, fistulated in the sigmoid colon. Finally, the colonoscopy confirmed this hypothesis with the visualization of a sigmoid fistula located 15cm from the anal margin in connection with a cavity containing hairs. Biopsies revealed a squamous cell carcinoma. After a negative extension assessment, a posterior pelvicotomy was performed with pelvic and lumbo-aortic lymph node dissection. The pathological diagnosis was an ovarian pT4N1R0 squamous cell carcinoma. An adjuvant chemotherapy is still in progress.

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Abbreviations

MCT: mature cystic teratoma; ODC: ovarian dermoid cyst; GCD: giant colonic diverticulum; SCC: squamous cell carcinoma; MRI: magnetic resonance imaging

Introduction

Mature cyst teratoma (MCT) or ovarian dermoid cysts (ODC) is the most frequent of benign ovarian tumors (60%) that mainly affect young women. On radiological procedures (CT-scan, MRI), the detection of an ovarian cystic tumor with fat components and calcifications confirms the diagnosis. However, some radiological presentations are less typical with misleading presentations that may suggest other diagnoses. Although benign in their current form, the risk of malignant transformation of the MCT is not zero, especially in perimenopausal women. The most frequent histological type encountered in cases of malignant transformation is squamous cell carcinoma (SCC). Its therapeutic management leads to surgical resection and chemotherapy.

Case Presentation

A 68-year-old patient was admitted to our department initially for suspicion of a complicated sigmoid diverticulitis. Her medical

history included osteoporosis and appendectomy. Following hypogastric abdominal pain for past 6 months, an abdomino-pelvic CT-scan showed a latero-sigmoid giant cyst with hydroaeric level and perisigmoid infiltration; giant colonic diverticulum (GCD) was mentioned. A pelvic MRI suggested a colonic-cystic fistula with a thickening and a peri-sigmoid infiltration and a left ovarian cyst 8 cm in diameter containing fat and hydro-aeric level, in favor of a mature cystic teratoma (MCT). The general condition was impaired with a weight loss of 4 kg (50 kg, 1m65). On clinical examination, abdominal pain was localized to the left iliac fossa and a fixed mass was palpated at the left pelvic level during the vaginal touch. After a new imagery review, the final hypothesis was that of a MCT complicated with a fistula between the sigmoid colon. Colonoscopy confirmed the diagnosis of fistula with a budding and an infiltrating lesion at 15 cm of the anal margin, in relation with a cavity containing hairs (Image 1). The biopsies of the upper rectum were characterized by a well differentiated squamous cell carcinoma (SCC), keratinizing and infiltrating, related to a MCT with a malignant transformation. After a negative extension assessment, a posterior pelvicotomy was performed by laparotomy and consisted of a left colectomy with stapling of the rectal stump and the confection of a left iliac colostomy,

a hysterectomy and bilateral adnexectomy, omentectomy, and pelvic and lumbar-aortic dissection. Two JJ probes were also set up in view of significant bilateral ureterolysis. The postoperative course was marked by an E. coli urinary tract infection healed with antibiotherapy. The patient was discharged on the 17th postoperative day. The histological examination confirmed a SCC related to a malignant transformation of a MCT, infiltrating the posterior walls of the uterus and cervix as well as the rectal wall and presenting lymphatic metastasis (2N + / 64N); either a pT4N1 lesion stage. The patient is currently on adjuvant chemotherapy.

Discussion

The case report describes an uncommon differential diagnosis of a MCT raising suspicion of a GCD localized on the sigmoid. Indeed, on the CT-scan, the aeric cyst and peri-sigmoid infiltration were initially confused with a giant diverticulum, compatible with painful symptomatology. GCD is a rare complication of diverticular disease, characterized by a diverticulum larger than 4 cm. The diagnosis is based on CT scan showing a gas-filled structure communicating with the adjacent colon with a thin diverticular wall without enhancing after injection of contrast [1]. The clinical presentation can be misleading, with symptoms that could suggest another digestive pathology and, like any other diverticulum, GCD can get complicated. Thus, cases of volvulus, abscess, perforation or bleeding have already been described in the literature [2-6]. Due to the associated risk of colon cancer (estimated at 2%), a colonoscopy should be performed. To avoid complications and with low morbidity and mortality rates, the colonic surgical resection (sigmoidectomy) with en-bloc resection of the diverticulum is the gold standard [7].

Mature cystic teratoma (MCT) or ovarian dermoid cysts (ODC) are the most common benign ovarian tumors, accounting for about 20-30% of all ovarian tumors developed from the germ cell line. They are most often diagnosed in young women (20-30 years), with few nonspecific symptoms (pelvic pain, pelvic mass, urinary symptoms). They are sometimes discovered fortuitously on a imaging procedure. Ultrasound can show an hypoechogenic cystic mass with an echogenic nodule in the wall (Rokitansky nodule), with, sometimes the presence of a lipid-liquid level [8,9]. In CT-scan, the presence of a level between the fat component and the liquid contained in the cyst is pathognomonic. Most often, MCT contains both fat, calcifications and tissue structures. In MRI, the fat saturated sequences are important, differentiating the hyperintense signal T1 from the fat component of a bleeding.

The main complications of MCT are rupture, torsion and malignant transformation [11]. Their risk of malignant transformation is rare, occurring in 1-2% of cases [12,13]. The clinical symptoms associated with malignant transformation are not specific, most often hypogastric pain or the discovery of a pelvic mass. However, this risk of transformation is more frequent in perimenopausal women (>50 years), when the lesion measures more than 10 cm and may be suspected with a high serum SCC antigen level [14]. SCC is the most frequent malignancy arising from MCT (80-90%), followed by adenocarcinoma (5%) and sarcoma (5%) [10].

On MRI, MCTs with malignant transformation tend to have a solid component and a transmural extension. In the case of advanced forms, there may be contact with others adjacent pelvic organs, or even, as in our case report, a digestive fistula with the sigmoid [15]. To our knowledge, this is the second case, after that of Min et al describing a sigmoid fistula complicating an ovarian SCC arising in a MCT. Other cases of SCC with digestive fistulas, with the small bowel have already been reported [12,16,17].

Although laparoscopy holds a major place in gynecological surgery and to avoid the risk of dissemination and spillage of cyst content; laparotomy is still recommended for SCC surgery [18,19]. A complete surgical resection (hysterectomy, bilateral salpingo-oophorectomy) with lymphadenectomy followed by adjuvant paclitaxel/carboplatin-based chemotherapy is the key to treatment [20].

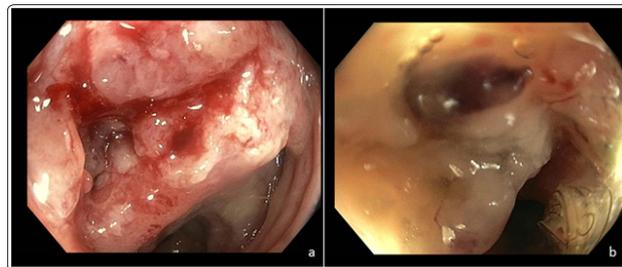


Figure 1: Colonoscopy showing, at 15 cm from the anal margin, a large bulging and infiltrating lesion (a) communicating with a cavity containing hairs (b)

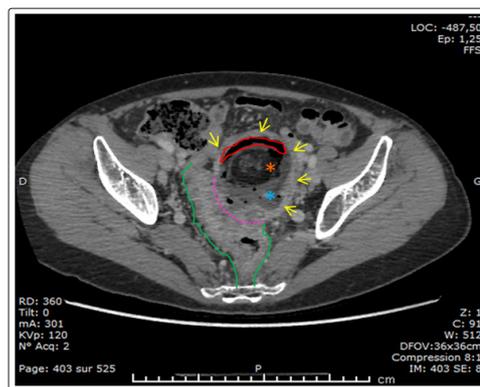


Figure 2: Axial section of an injected pelvic CT-scan showing thickened and infiltrated sigmoid walls (green lines), with a zone of continuity with a left ovarian cyst about 6 cm (pink dotted line). The left ovarian cyst, 8 cm in diameter, presents thickened walls (yellow arrows) and heterogeneous contents in favor of a dermoid-cyst : fat and sebum (orange asterisk), upper aeric level (red line) and liquid with air bubbles (blue asterisk) suspicious of a fistula with intra-cystic abscess.

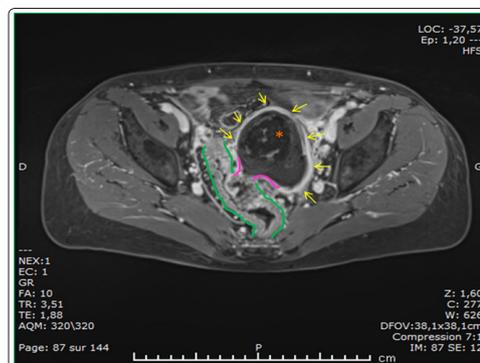


Figure 3: Axial section of a pelvic MRI in T1 sequence with gadolinium injection. Yellow arrows show the thickened walls of the left ovarian dermoid cyst containing fat and sebum (orange asterisk); green lines show the infiltrated and thickened sigmoid wall and the pink lines the fistula between these two organs.

Conclusion

This case report describes an unusual presentation of MCT with malignant transformation and sigmoid fistula. The initial doubt with a giant colonic diverticulum diagnosis, was eliminated after

an exhaustive assessment and a multidisciplinary management, thus radically changing the therapeutic strategy.

Declarations

Authors' contributions

LM collected data and drafted the manuscript. FP participated in the proofreading and interpretation of radiological procedures. AL and AC performed the surgical procedure and reviewed the manuscript. RF reviewed the manuscript. All authors read and approved the final manuscript.

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Not applicable.

Competing interests

The authors declare that they have no competing interests

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