GLIOMATOSIS PERITONEI IN A PATIENT WITH A BENIGN OVARIAN CYST

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Introduction:

Gliomatosis Peritonei (GP) is a rare occurrence and as such the pathogenesis is not yet fully understood. It is characterized by the implantation of glial tissue on the peritoneal surfaces and can be associated with solid ovarian teratomas. Its occurrence with mature cystic teratomas is rare with only about one hundred cases in the literature worldwide. We present what may be the first case in the English-Speaking Caribbean.

Case History:

A 50-year-old, para 4 female with no known medical conditions presented to the Accident and Emergency department with a two-week history of generalized abdominal pain and swelling. She also complained of significant weight loss. There was no pertinent past medical, surgical or family history. On examination, she was found to be cachexic with temporal wasting and had a grossly distended abdomen with ascites and a large abdominopelvic mass. Although her blood count, kidney and liver functions were normal, her tumour markers were elevated [Ca125 – 500 U/ml (0.0-35.0 U/ml) and Ca19.9 – 10,000 U/ml (0.0-27.0 U/ml)].

Imaging revealed a large, well-defined heterogeneous mass containing fat, calcific foci and enhancing soft tissue components measuring 17.7cm x 20.5cm x 23.4cm possibly from the left adnexa with ascites and peritoneal thickening (see figures 1 and 2). There was no lymphadenopathy. An ascitic tap revealed no malignant cells, however, the fluid was very milky and turbid in appearance. The patient was counselled and optimised for debulking surgery.

Figure 1: Axial image of contrast-enhanced computed tomography (CECT) of the abdomen and pelvis showing the heterogeneous adnexal lesion comprising of fat and soft tissue components with surrounding ascitic fluid

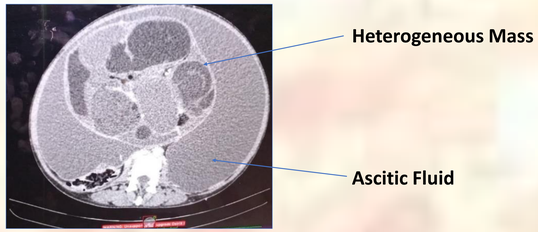
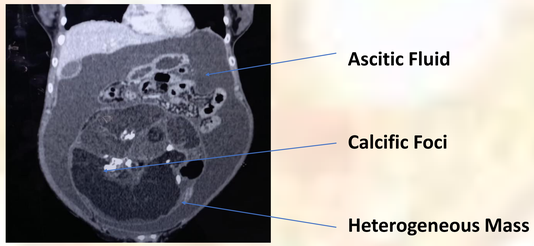


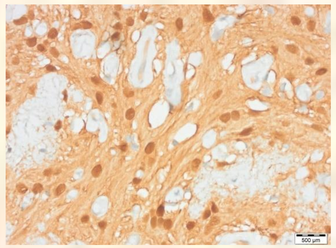
Figure 2: Coronal image of CECT of the abdomen and pelvis showing the heterogeneous adnexal lesion comprising of fat, calcific foci and soft tissue components with surrounding large volume ascitic fluid.



She underwent laparotomy with total abdominal hysterectomy, bilateral salpingo-oophorectomy with excision of the large abdominopelvic mass, omentectomy, appendectomy, and extensive adhesiolysis as well as ascitic fluid cytology. There were extensive adhesions involving loops of both small and large bowels as well as omentum, to the unruptured abdominopelvic mass which extended up to the xiphoid process. The small and large bowels seemed to form a congealed mass pushed up to the left side of the abdomen. There was no macroscopic residual disease post debulking surgery. She recovered well postoperatively and was subsequently discharged.

Histologic analysis revealed a mature cystic teratoma with no evidence of immature elements or malignancy. The omentum, however, showed implants of mature glial tissue with no immature components being identified. The peritoneal fluid was also positive for glial cells. Both findings were suggestive of gliomatosis peritonei and were confirmed by S-100 immunohistochemical stain (see figure 3).

Figure 3: S-100 immunohistochemical stain confirming the presence of glial tissue in the omentum.



One year post-procedure, the patient remained well with no abnormal symptoms. All her tumour markers have normalised and her imaging showed no recurrence.

Discussion:

Gliomatosis peritonei (GP) is a rare condition characterised by the presence of benign glial implants in the peritoneum, omentum and lymph nodes. Whilst it is usually associated with immature ovarian teratoma, rarely, it can be seen with a mature teratoma as in our case [1]. Accurately accounting for its incidence is difficult as only approximately 100 cases have been reported worldwide [1].

The exact pathogenesis of GP is uncertain. However, from some case reports, there are currently two working theories about its aetiology. The first is related to spread through ovarian capsular flaws and angiolymphatic channel dissemination, whilst the second suggests that pluripotent stem cells in the peritoneum or underlying mesenchyme undergo glial metaplasia [1]. Intra-operatively, our patient had extensive intra-abdominal adhesions. Given that she had no previous surgeries or pelvic infection, as well as there being the apparent resolution of GP following surgery, favours the first theory of spontaneous capsular leak or rupture. Capsular leaks result in the release of an exudate rich in pro-inflammatory cytokines which leads to adhesion formation.

Although imaging studies can aid in identifying GP, its appearance however, can mimic peritoneal tuberculosis or a metastatic process. Glial deposits are usually small and can even be sub-centimetre, hence it can be missed on imaging studies [2]. Therefore, the definitive diagnosis of GP is based on histopathological analysis, where the specimen is generally positive for markers like glial fibrillary acidic protein (which determines the maturity of the tissue), S-100 protein and neuron-specific enolase [3]. In our case, the S-100 protein immunohistochemical stain was used to aid in the diagnosis of the condition.

As GP is classified as a grade 0 teratoma according to the WHO criteria [4], the prognosis is usually good. Surgical resection is usually the treatment of choice, although complete resection is near impossible when the disease process is quite extensive. Studies have shown that residual peritoneal disease can be asymptomatic and quiescent over a long period of time or may even spontaneously resolve [2,5]. The treatment decision is usually based on the maturity of the primary tumour and the glial tissue implants. As long as the implants have been thoroughly sampled and are all mature, a relatively conservative approach to follow-up can be undertaken. However, if immature glial tissue or other teratomatous components, or both, are found in the peritoneum or omentum, the therapy should be similar to that for metastatic ovarian teratoma [6].

Even though GP has such a generally good prognosis, long-term surveillance in the setting of mature glial implants is advised as there is the potential for malignant transformation of the glial components long after the initial surgery [7]. There is no consensus as to the follow-up care in patients with GP [1] and as such, our patient will continue to have regular follow-up.

Conclusion:

GP, although a rare entity, has excellent prognosis. The mainstay of treatment is surgical excision followed by close clinical follow-up.

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