LETTER TO THE EDITOR

Incidental finding of pseudomyxoma peritonei in a deceased organ donor: implications on organ donation

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Pseudomyxoma peritonei (PP) is a rare disease characterized by the deposits of mucinous material in the peritoneal cavity secondary to rupture or metastases of a primary neoplasm. The primary tumour can be benign or malignant, commonly of the appendix or ovary. There is no written literature on the implication of an incidental finding of PP on organ donation. We have recently discovered one such case at the time of organ retrieval in a deceased nonheart beating donor (NHBD).

The donor was a 34-year-old Caucasian male who had sustained a head injury secondary to a road traffic accident. He fulfilled criteria for Maastricht category III controlled NHBD. He had no significant co-morbidity. There was no contraindication to donation on standard donor screening protocol. The retrieval was performed through a midline laparotomy. A gelatinous substance was noted on the peritoneal surfaces of the right lobe of the liver, right hemi-diaphragm and right paracolic gutter. The appendix was enclosed in a mass of gelatinous substance. On closer inspection, the gelatinous material was adhered to the capsule of the liver and was difficult to peel off (Fig. 1). The appendix was thickened in the middle giving it a fusiform appearance with a small perforation. The gelatinous material from the surface of the liver and the appendix was removed. Frozen section histology was requested of the appendix. The rest of the retrieval process was performed in the usual manner and the liver and kidneys were removed for possible transplant pending histology results.

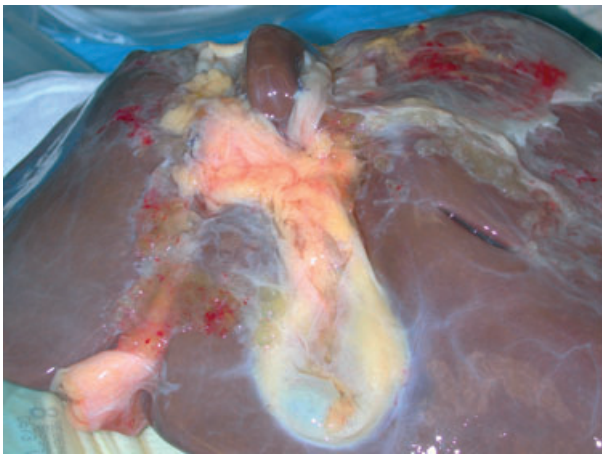

Figure 1 Operative photograph showing gelatinous material adherent to the under surface of the liver.

The frozen section of the appendix suggested an adenoma with pseudo-stratification and pleomorphism. Mucin material was seen to dissect through the wall with foci of necrosis and dystrophic calcification. The appearance suggested adenomucinosis (PP). The presence of malignancy could not be excluded on frozen section. Because of the short cold ischaemic tolerance of the liver, it was decided not to proceed with liver transplantation. A decision to transplant the kidneys was postponed until the definitive histology from paraffin sections was available.

The paraffin section of the appendix revealed similar findings. However, definite invasion by epithelial neoplastic cells was not demonstrated. Most of the mucosa was attenuated and denuded and the wall was dissected by pools of mucin with associated necrosis and dystrophic calcification. The gelatinous material from around the liver was microscopically shown to be a pool of mucin with small fragments of adenomatous gland in some of them. At this juncture we sought advice from a national expert who had experience in dealing with PP. The advice was not to use the abdominal organs for transplant even if the primary tumour was benign.

Pseudomyxoma peritonei, otherwise known as 'jelly belly', was first described over a century ago [1]. It is characterized by extensive mucinous implants on the omentum and peritoneum and production of gelatinous ascites in the peritoneal cavity. Morphological and genetic studies have shown that the disease most commonly originates from the appendix which ruptures resulting in the release of mucous containing epithelial cells into the peritoneal cavity [2,3]. This collects at sites within the abdominal and pelvic cavity; a process described as the 'redistribution phenomenon' [4]. Many other primary tumours have been reported to be associated with PP [5] demonstrating a marked heterogeneity in the pathology of this disease. Furthermore, there is little consensus on

whether PP should be classified as malignant or not, particularly to differentiate it from carcinomatosis peritonei secondary to high-grade mucinous carcinoma. Irrespective of the histological grade, PP behaves aggressively by local growth and dissemination on to the peritoneal surfaces. Vascular invasion and/or blood or lymph borne metastasis is extremely rare.

There continues to be debate regarding treatment of this condition. The Sugarbaker procedure was described as a potential treatment option [6]. This involves extensive debulking surgery with intraperitoneal and systemic chemotherapy. It is associated with a 2 and 5 year survival rate of 95% and 70%, respectively [7]. However, there remains a large morbidity associated with this treatment. Recurrence is almost universal requiring repeated surgery to debulk the growth and or chemotherapy. A recent review article concluded the decision making about radical treatment for a rare condition is difficult especially when evidence on clinical and cost effectiveness is not so clear cut. Currently, guidance issued by the National Institute for Clinical Excellence (UK) states that current evidence of efficacy and safety of complete cytoreduction does not appear adequate for this procedure to be used in the National Health Service (NHS) outside centres specifically funded for such treatment by the National Specialist Commissioning Advisory Group (NSCAG) [8].

Pseudomyxoma peritonei remains rare in the UK with reported incidence of 50 new cases a year. Exposure to this condition remains limited among surgeons, particularly transplant surgeons. This is further complicated by the fact that establishing the nature (benign or malignant) is not always possible by conventional histological technique in the time that is available. The natural history of the disease irrespective of its benign or malignant nature and histological grade is aggressive. Recurrence is almost universal after resection or cytoreduction. In a transplant setting, it is deemed to be an even higher risk to transplant such tissue combined with the immunosuppressive therapy required postorgan transplant. Organs such as the liver, small bowel and pancreas will be unsafe to transplant even after excision of all the visible surface lesions. The kidneys being a retroperitoneal structure and the fact that all the peritoneal covering is removed prior to transplant, can potentially be used. However, the approach to the kidney is transperitoneal and spillage and implantation of cells cannot be excluded even with utmost care. Therefore, safe advice will be not to use any abdominal organ for transplantation from donors with PP. Awareness of this condition among the retrieving surgeons is therefore crucial to decision making.

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