PSEUDOMYXOMA PERITONEI

Patient Information Leaflet

Partners: Brendan J Moran Tom D Cecil Steve J Arnold Faheez Mohamed Arcot K Venkatasubramaniam

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Pseudomyxoma peritonei is a rare disease of the abdominal or peritoneal cavity. The majority of cases result from a ruptured mucus secreting adenoma of the appendix. More rarely, the condition arises in other parts of the bowel, in the ovary in females and in rare cases, in other organs such as the bladder. The disease is due to the presence of a large amount of mucinous fluid with the consistency of jelly in the abdominal cavity, and is often commonly referred to as 'Jelly Belly'. The mucus is produced by cells that have spread and grown on the peritoneum which lines the abdominal cavity.

Pseudomyxoma peritonei is often referred to as being a 'borderline malignant' condition. The characteristics of a malignancy are that it spreads via the lymphatic system to the lymph nodes, and by the blood vessels (vascular spread), thus reaching the liver, lungs, brain and other tissues. Pseudomyxoma peritonei spreads only within the abdominal cavity in the majority of cases and does not spread via the lymphatics or the blood stream. However, it is not completely benign, as it continues to grow and eventually takes over the peritoneal cavity. If untreated, it will result in compression of the abdominal organs, making normal nutrition no longer possible. The complications of malnutrition (predominantly infection such as pneumonia), and complications following surgical treatment of the disease are common.

Pseudomyxoma peritonei of appendix origin

Pseudomyxoma peritonei of appendix origin is said to have an incidence of around one per million per year. Common presentations of the disease are abdominal distension, mucus in a hernia sac, perforated appendix, or an ovarian mass in females. Occasionally, it is found incidentally at laparotomy (explorative abdominal operation). The sequence of events is thought to be rupture of a mucus-secreting adenoma of the appendix with release of viable (live) adenoma cells into the peritoneal cavity. These cells spread widely within the peritoneal cavity. They become attached to the peritoneum and release mucus, producing mucinous ascites (fluid and jelly). The main concentrations of cells follow the flow of peritoneal fluid and tend to accumulate at particular sites within the abdominal cavity.

Mucinous adenocarcinoma

There is a wide spectrum of disease, varying from the benign at one end of the spectrum to mucinous adenocarcinoma at the other. Thus, many patients who present with mucinous ascites suggestive of pseudomyxoma peritonei of appendix origin turn out to have mucinous adenocarcinoma. Histology of the two conditions is quite similar in that there are atypical (abnormal) cells with large mucinous pools. Often the differentiation is only possible by the clinical behaviour of the disease and by detailed pathology of large volumes of resected specimens. The condition is commonly confused with ovarian cancer in women, and particularly with mucinous low malignant potential ovarian tumours. A small ruptured tumour of the appendix can result in very large ovarian tumours and it is often quite difficult to rationalise that a large ovarian tumour can be secondary to a small ruptured tumour of the appendix.

Treatment Options

For most tumours there are four options which can be considered.

1. **Watch and wait**

2. **Radiotherapy**
Radiotherapy does not have a place in the management of pseudomyxoma peritonei, as it would be impossible to apply radiotherapy to a large area without causing serious damage to the abdominal organs.

3. **Chemotherapy**
The commonly used forms of chemotherapy (oral or intravenous) have very little role at the benign end of the spectrum. This is due to the fact that the disease is of borderline malignancy and has a very poor blood supply, so that chemotherapy does not gain access to the cells. All chemotherapy treatment relies on a balance between the benefits and risks. For a low grade tumour the risks of treatment far outweigh the benefits, and therefore the majority of oncologists (chemotherapy specialists) consider that chemotherapy has no place in the management of early pseudomyxoma peritonei. However intestinal type chemotherapy sometimes has beneficial effects if the tumour is a mucinous adenocarcinoma.

4. Surgery - broadly of two types:

Debulking

The common surgical approach is debulking to remove as much of the tumour as possible, and generally includes removal of the uterus and ovaries in the female and often the right colon and the omentum. Disease recurrence is almost inevitable due to residual and recurrent disease around the peritoneal cavity. Repeat debulking surgery may be possible on a number of occasions, but each attempt becomes more difficult and dangerous. The small bowel becomes increasingly involved due to adhesions following prior surgery and eventually surgery is impossible and is fraught with severe complications such as small bowel fistulae.

Complete cytoreduction

Complete cytoreduction (complete tumour removal) is a technique developed and popularised by Professor Paul Sugarbaker at the Washington Cancer Centre. Average operating time for what is called a 'major peritonectomy' is ten hours. The operation comprises a number of different procedures, namely:

- Right hemicolectomy
- Greater omentectomy
- Splenectomy
- Cholecystectomy
- Lesser omentectomy
- Pelvic peritonectomy, which sometimes includes the rectum by anterior resection and in the female includes removal of the ovaries and uterus
- Stripping of the peritoneum from the left hemidiaphragm
- Stripping of the peritoneum from the right hemidiaphragm
- Stripping of disease from the surface of the liver

An important factor at surgery is the involvement of the small bowel. In general the small bowel is not grossly involved due to small bowel peristalsis (normal movement of the bowel) being relatively protective against tumour implantation and growth.
The best mechanism for determining involvement of the small bowel prior to surgery at present is a CT scan with a large volume of oral contrast to outline the small bowel.

On the CT scan it is possible to see tumour displacing the small bowel and in the small bowel mesentery - usually a poor prognostic factor. Most cases have some degree of small bowel involvement but it is usually possible to deal with limited small bowel disease. If all disease can be removed, heated Mitomycin C is given directly into the peritoneal cavity at the time of operation at a temperature of approximately 400C. Intraperitoneal 5FU is often given for four days post-operatively.

Suitability for surgery

Not every case is suitable for surgery for a number of reasons - in particular, the extent or distribution of the disease, or fitness to withstand major high risk surgery. Occasionally, it is not possible to determine the extent of disease adequately until the abdomen has been opened, and therefore it is never possible to guarantee that complete tumour removal will be achievable.

Post-operative Mortality and Morbidity

Complete cytoreduction carries a mortality risk of 3-5%, which means 1/30 to 1/20 patients die as a direct result of surgical complications. Basingstoke currently has a 1-2% mortality rate. The main complications are cardio-respiratory (lung infections and heart failure). There is also a risk of clots in the main leg veins, which can result in pulmonary embolus.

Surgery also has significant morbidity (serious complications) of around 30%. Approximately 20% (1 in 5) patients require further surgery to deal with the complications of the primary operation during the same admission. Approximately 20% of patients require a stoma and in half of those the stoma will be permanent. A permanent stoma is required if all or most of the colon has to be removed. A temporary stoma is usually used when the rectum has to be removed and the join, although appearing intact at the time of surgery, has a very high risk of leakage due to the particular position of the anastomosis or join, and the fact that intraperitoneal chemotherapy is used. The temporary stoma is usually closed between 3 and 6 months after the primary operation.

Most patients spend about 3 days in intensive care, and the average hospital stay is 21 days. All patients require feeding by intravenous or total parenteral nutrition (TPN) for an average of 14 days. Patients can take a minimum of six months until they are fully active and able to return to work. Symptoms can affect the ability to undertake all usual activities whilst recovering from what is very major surgery.

If complete tumour removal has been possible, intraperitoneal chemotherapy has been given and the tumour is at the benign end of the spectrum, 50-80% will have ten year disease free survival.

ALL CORRESPONDENCE AND APPOINTMENTS

The Hampshire Clinic Basing Road Basingstoke Hampshire RG24 7AL
T: 01256 354747 F: 01256 818005
E: info@basingstokecolorectal.co.uk W: www.basingstokecolorectal.co.uk