Malignant soft tissue tumours of the trunk and retroperitoneum

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Malignant soft tissue tumours of the trunk, retroperitoneum and viscera account for nearly 40% of all soft tissue malignancies. The wide variety of anatomical sites of origin, the diverse histological subtypes and the rarity of these tumours has made meaningful study of these lesions difficult. There is no doubt, as experience in the management of these lesions has improved, unique features related to site of origin, pathological subtype, tumour biology, response to treatment etc, have become apparent. For most patients the outlook remains poor. Surgery remains the principal form of treatment.

Retroperitoneal soft tissue sarcomas

Soft tissue sarcomas arising in the retroperitoneum account for approximately 15% of all soft tissue tumours (Storm and Mahvi 1991). The advanced size of most tumours at the time of diagnosis and their proximity to major retroperitoneal and intra abdominal structures further complicates the management of this disease. The poor results from surgical management of these lesions is due to the lack of fascial planes which tend to contain sarcomas elsewhere in the body and the difficulty in obtaining wide margins of excision. It may be difficult to distinguish tumours arising from retroperitoneal structures from soft tissue tumours arising from other structures within the abdominal cavity particularly when the tumour is large. Primary intra abdominal sarcomas are rare compared to retroperitoneal soft tissue tumours.

The median age of presentation is approximately 55 years and both sexes are equally effected. Patients with retroperitoneal masses characteristically present late. Many patients in retrospect describe trivial symptoms present for many months prior to diagnosis. Eventually most patients complain of abdominal and or back pain of variable severity. Urgent symptoms such as bowel obstruction or urinary tract obstruction occur in less than 5% of patients. An abdominal mass is palpable in the three quaters of patients. Weight loss is an uncommon manifestation of retroperitoneal tumours (7%). A variety of unusual symptoms such as neurological symptoms due to nerve invasion, leg oedema or varicocele secondary to venous obstruction and hypoglycaemia due to production of insulin like factors by large retroperitoneal fibrosarcomas have all been reported (Jaques et al. 1988).

The diagnosis is most often suspected on the basis of CT or MRI. MRI is the examination of choice because it provides improved definition of anatomical structures and the tumour. Multi planar reconstruction which may help in the planning of surgery is available as is magnetic resonance angiography if information concerning vascular anatomy is necessary. The presence of intra abdominal spread or liver metastases should be sought . CT scanning of the lungs is mandatory to exclude metastatic disease. Approximately 20% of patients presenting with retroperitoneal soft tissue tumours are found to have evidence of metastatic disease at the time of initial work up.

Approximately 80% of retroperitoneal masses diagnosed on CT or MRI are found to be malignant while the remaining 20% are benign. Slightly more than half of the malignant lesions turn out to be sarcomas. Other common retroperitoneal malignancies include lymphomas and urogenital malignancies.

Despite the advances in imaging it is generally not possible to distinguish between benign and malignant lesions and between sarcomas and other malignancies. Histological confirmation of the diagnosis is therefore essential in planning management. Biopsy potentially poses the major risk of tumour contamination of the abdominal cavity. Transperitoneal needle biopsy is to be avoided at all costs because of the enormous potential for seeding the abdominal cavity. Laparoscopic biopsy suffers from the same problem and should also be avoided. Needle biopsy of retroperitoneal tumours from the back avoids the problem of uncontrolled contamination of the abdominal cavity but the potential for seeding of the needle track needs to be considered. Our preference is to work the patient up for a resection and at the time of operation perform an open biopsy through a limited abdominal

incision with a view to proceeding if the diagnosis of sarcoma is confirmed on frozen section. A representative piece of tumour can be sampled with strict attention to preventing tumour contamination of the peritoneal cavity. The considerable vascularity of many retroperitoneal sarcomas, their friable consistency and the predilection for cystic necrosis in large lesions can make safe open biopsy difficult. Frozen section diagnosis of retroperitoneal soft tissue tumours can be difficult and occasionally it may be necessary to secure the biopsy site, terminate the procedure and await the definitive histology report.

Liposarcomas are the commonest pathological subtype and account for nearly half of retroperitoneal sarcomas. Fibrosarcoma is commonly noted in older series or series which have accumulated patients over several decades. Many of these tumours would now be classified as liposarcomas. The diagnosis of malignant fibrous histiocytoma (MFH) has become more common in recent years. Nevertheless liposarcoma remains by far the most common subtype. Approximately 60% of liposarcomas are low grade and 40% high grade. Leiomyosarcoma, predominantly high grade, is the next most common pathology accounting for at least one quarter. MFH and sarcomas of neural origin make up most of the remainder apart from a small group of undifferentiated, unclassifiable tumours. Identification of sub-type and tumour grade is important in indicating the likely natural history of the tumour. Tumour size, and histological subtype (liposarcoma favourable, leiomyosarcoma unfavourable) have variously been found to be related to survival by many groups. Complete resection versus incomplete resection and probably tumour grade however is the only factor that reliably predicts outcome (Karakousis et al. 1985, Catton et al. 1994, Glenn et al. 1984, Jaques et al. 1989, Catton et al. 1994, Singer et al. 1995).

Surgical management can be extremely challenging because of the extensive size of tumours at presentation and their relationship to other organs and the major neurovascular structures of the retropeitoneum. Complete resection is possible in approximately 50% of cases (Table). Despite improvements in preoperative assessment with MRI operability can only be accurately determined at laparotomy. If the patient is fit for operation then our policy is to proceed to laparotomy. Most retroperitoneal tumours involve either side of the abdominal cavity arising from the vertebral gutters. Central lesions are uncommon except when tumours have attained massive size or involve the pelvis. A thorough laparotomy should be performed to exclude metastatic disease particularly in the liver. Intra abdominal satellite nodules may be found particularly if the operation is for recurrence. They are resected with an appropriate margin. Unfortunately the chances of subsequent intra abdominal recurrence are high. The liver should be inspected closely. Liver metastases at the time of first presentation are uncommon. Generally we would not perform a liver resection at the time of a definitive tumour resection unless it was a limited low morbidity procedure.

Tumour resectability is assessed at this stage. Involvement by tumour of adjacent structures is common and some form of en bloc excision with removal of adjacent organs is necessary in up to 75% of patients. Storm and Mahvi (1991) emphasise the importance of a bi-manual examination by entering a plane adjacent to the vertebral body and lateral to the great vessels of the abdomen. The other hand is placed deep to the tumour through an incision in the lateral abdominal wall well outside the tumour. An assessment of fixation of the tumour can then be made. Tumours are inoperable if there is extensive fixation to the posterior abdominal wall musculature. Experience is required in assessing the degree of fixation and if resection can be safely undertaken.

It is commonly claimed that soft tissue tumours do not infiltrate or invade structures and therefore major neurovascular structures may be preserved during resection of large tumours. They frequently encircle and entrap major vessels particularly in the pelvis, severely limiting resectability. While mobilisation of arteries away from sarcomas is generally possible, major veins are far more vulnerable to inoperable tumour encasement. Large tumours extending into the pelvis provide some of the most difficult surgical problems. For upper abdominal lesions a thoraco abdominal incision may be necessary. Sarcomas involving the mesentery provide particular problems related to safeguarding the integrity of the mesenteric vessels.

Throughout any operative procedure great care must be taken while dissecting the tumour to avoid tumour spillage. Many large sarcomas are covered in numerous delicate superficial vessels which are easily damaged. The tumour tissue is often quite friable and if central tumour necrosis has occurred, rupture of the

Retroperitoneal sarcoma

	No.	Completely resected (%)	5-year survival
Karakousis et al. 1985	68	40	64
Jacques et al. 1990	114	59	60
Catton et al. 1994	45	–	55

tumour with consequent tumour dissemination can occur.

Less than complete resection only is possible in approximately 20% of cases (Storm and Mahvi 1991). Incomplete resection appears to have some short term effect over biopsy alone but this is short lived and any effect is lost by 5 years (Storm and Mahvi 1991, Jaques et al 1989). Nevertheless subtotal resection potentially provides improved symptom control for a variable period and on that basis is worthwhile. The benefits of partial resection can be especially gratifying with low grade tumours providing symptom control for years. Overall survival in most reported series is approximately 55% at five years with a range from 30-75% (Table 1). Recurrence after 5 years is not uncommon and in one series the 2 year survival of 34% had fallen to 17% at 5 years and 8 % at 10 years (Storm and Mahvi 1991). It is not uncommon for low grade tumours to recur after many years.

Local or intra abdominal recurrence is the commonest site of failure and will occur in the majority of patients who eventually succumb to the disease. In three quarters of cases intra abdominal recurrence is the first site of recurrence. The lung and liver are the major sites of blood borne metastasis. Most high grade tumours which recur intra abdominally do so within the first two to three years (Storm and Mahvi 1991). Jaques et al (1989) report a resection rate of 44% with a 41 month median survival time for surgical management of recurrent intra abdominal disease. The majority of patients who do develop intra abdominal recurrence will do so again and usually at a shorter time period than before development of the first recurrence. The phenomenon of deteriorating grade or dedifferentiation is a feature of low grade liposarcomas of the retroperitoneum and to lesser extent leiomyosarcomas and goes some way to explaining the more aggressive behaviour of recurrences and the increasingly shorter times to subsequent recurrence.

The symptoms associated with local recurrence are similar to initial presentation with the widespread nature of intra abdominal disease associated with a higher incidence of abdominal distension, early saiety, loss of weight and abdominal discomfort. These symptoms can often be palliated for a time with incomplete resections.

The use of adjuvant radiotherapy is not justified based on the results to date. The total radiation dose is limited by the toxic dose to adjacent organs particularly bowel. Glenn et al. (1984) who used a total dose of 55 grey noted a very high rate of bowel complications without any improvement in survival. Unfortunately intraoperative radiotherapy which offers the possibility of rapidly treating the tumour bed with high dose radiotherapy without the concerns of adjacent tissue damage by radiation has not been found to be effective (Willett et al. 1991).

Similarly, chemotherapy has not been shown to improve survival in patients with retroperitoneal soft tissue tumours (Storm et al, Glenn et al.1985).

Soft tissue sarcomas of the abdominal wall

Soft tissue sarcomas of the abdominal wall are rare. Weinstein and Shiu (1989) reported a total of 55 presented to Memorial Sloan Kettering during the period 1949–1982. In this series, desmoids accounted for one third of tumours. Rabdomyosarcoma, MFH, synovial cell sarcoma and liposarcoma were responsible for the remainder. Patients usually present with a palpable mass which may initially be diagnosed as a benign tumour, ie. lipoma, or hernia until a biopsy is performed. Principles of management are similar to extremity soft tissue sarcomas. The lesion should be widely excised and in most cases this will necessitate full thickness abdominal wall with in continuity of resection of any compromised intra abdominal viscera. In many cases the defect can be repaired primarily. Reconstruction of the abdominal wall with polypropylene mesh, particularly if there has been little in the way of skin and other soft tissue loss is the reconstructive technique preferred by many. Our preference, particularly for recurrent disease, or when post operative radiotherapy is planned is for a free tensor fascia lata flap.

Post operative radiotherapy is reserved for recurrent desmoid tumours, large primary abdominal wall sarcomas, particularly where there may be any compromise of margins, and recurrent abdominal wall sarcomas. High dose radiotherapy is not without problems particularly to underlying bowel and other intra abdominal organs which may limit the dose.

Five years survival for non desmoid sarcomas in the small series reported by Weinstein and Shiu (1989) was 22% at five years. Local control is possible in the majority of cases. The predominant site of distant failure was the lungs.

Soft tissue sarcomas of the chest wall

Soft tissue sarcomas of the chest wall are as uncommon as lesions involving the abdominal wall. Most patients present with an enlarging mass and only rarely is there evidence of intra thoracic involvement suggested by shortness of breath, pleuritic pain, haemoptysis, etc. Most chest wall tumours are malignant and most of these are metastatic lesions ie recurrent breast cancer, lung cancer, plasmocytoma. Desmoids of the

chest wall are relatively uncommon accounting for only 15% of lesions. Malignant peripheral nerve tumours, MFH, fibrosarcomas, liposarcomas and rhabdomyosarcomas account for most chest wall sarcomas (Bains et al. 1990). Management is predicated upon complete resection and this may require some form of chest wall reconstruction. A small defect can be repaired primarily. Larger defects may be managed with a free flap whilst extensive resections with instability of the chest wall have been managed with a combination of polypropelene mesh reinforced with methyl methacrylate (Faber et al. 1995). Radiotherapy is indicated for high grade lesions particularly when margins are compromised and for recurrent sarcomas. Outcome depends primarily on the tumour grade, the ability to obtain a sufficient margin and to a lesser extent tumour size. Survival is similar to abdominal wall sarcomas and is primarily related to grade and the completeness of the surgical excision.

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