the high level resistance. Removal of infected intravenous devices or other prosthesis must be considered and may be the only treatment available.

Infection control implications must not be overlooked and in our case it was fortunate that the patient was nursed in isolation since his admission for the bone marrow transplant. It remains to be seen whether standard infection control procedures will be sufficient to prevent spread of vancomycin-resistant enterococci. A review of the current use of vancomycin and cephalosporins may also need consideration.

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Intra-Abdominal Desmoplastic Small Round-Cell Tumour: Response to Multimodality Treatment

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Summary

A Malaysian case of this recently-described aggressive tumour is reported. A 16-year-old Malay boy presented with a large abdominal mass and iron deficiency anaemia. After incomplete resection of the mass at laparotomy, the residual disease showed response to chemotherapy. However, 'second-look laparotomy' showed nodules of residual disease in the liver. He underwent further chemotherapy and total abdominal radiotherapy. The patient remained in remission for 6 months, but relapsed and died 30 months after diagnosis. This tumour is responsive to aggressive multimodality therapy, but most patients die of their disease.

Key Words: Abdominal neoplasm, Combined modality therapy, Desmoplastic, Small cell tumour

Introduction

Intra-abdominal desmoplastic small-round cell tumour was first clearly separated from other small round-cell tumours (SRCT) of infancy and childhood in 1989[1], since when interest has increased rapidly, with most cases reported from the USA and Europe[2]. Outside these areas, two cases have been reported from Australia, and one from Hong Kong. Clinically, these highly aggressive tumours commonly affect adolescent males and present with masses in the pelvis or abdomen arising from the peritoneal surface[2]. Pathologically they show a nesting pattern of growth with an intense desmoplastic reaction and immunoreactivity for epithelial, neural and muscle markers. We report a Malaysian case: a boy who demonstrates the typical clinical features, a partial response to chemoradiotherapy but, ultimately, the poor prognosis of this tumour.

Case Report

A 16-year-old Malay boy presented with symptoms of anaemia for five months in May 1993. There had been fever, loss of appetite and marked loss of weight. Examination revealed a thin pale boy with a large, firm and mobile mass in the left upper quadrant of the abdomen. His temperature was 37.8°C, but there was no other abnormality on examination. Investigations showed him to have a severe iron deficiency anaemia (Hb 2.6 g/dl) and a raised alkaline phosphatase (398 iu/L). Ultrasound and CT scan showed a mobile mass encircling a loop of small bowel and multiple enlarged para-aortic lymph nodes. A fine needle aspiration of the mass was performed (see below). At laparotomy, a hard tumour measuring 10x10x10cm was found arising from the jejunal mesentery involving two adjacent loops of jejunum. Unresectable enlarged lymph nodes were noted in the mesentery and at its base. The primary tumour was resected in one piece together with 76 cm of jejunum and its mesentery.

After surgery, the patient felt much better, the fever settled and he put on weight. A repeat CT scan showed two hypodense lesions in the right lobe of the liver and enlarged matted lymph nodes in the region of the head of the pancreas. A chest X-ray, bone scan and bone marrow were normal. He underwent 7 cycles of chemotherapy as shown in Table I. Repeat CT scan at the end of this treatment showed complete resolution of the enlarged lymph nodes and no evidence of residual disease in the liver.

A second-look laparotomy was undertaken in February 1994, at which no residual disease was seen at the primary site or at the site of previous lymphadenopathy (random biopsies of mesenteric lymph nodes were negative). However, four tiny nodules were seen within the right lobe of the liver, which showed metastatic disease histologically. He thus underwent two further cycles of chemotherapy with some dose escalation (Table I), supported by prophylactic use of granulocyte-macrophage colony stimulating factor. Finally, in October 1994, he underwent total abdominal radiotherapy from both anterior and posterior fields (30Gy, with shielding of the liver and kidneys at 15Gy). In December 1994, 20 months after diagnosis, he was well, and abdominal CT scan showed no detectable disease. However, he began to show signs of progression in June 1995, with clinically and radiologically apparent metastatic disease in the liver. He died in November 1995, 30 months after diagnosis.

Histopathology

The fine needle aspiration from the abdominal mass showed malignant cells, the appearance of which was suggestive of rhabdomyosarcoma. Histological examination of the excised laparotomy specimen showed nests and sheets of tumour cells separated by desmoplastic stroma. One of five excised mesenteric lymph nodes was positive for tumour cells. Immunocytochemical studies were performed using an indirect immunoperoxidase technique. These showed the tumour cells to be positive for the following antigens: cytokeratin (AE1 and AE3), neurone-specific enolase, synaptophysin, desmin (focally positive) and myoglobin, S-100, α-fetoprotein, α_1-antitrypsin and α_1-chymotypsin. The tumour cells were negative for carcino-embryonic antigen and chromogranin.

Discussion

Desmoplastic small round-cell tumour is a recently recognised tumour affecting mainly children and young adults of male sex[1]. Its distinction from other SRCT

Table I
Treatment schedule for this patient suffering from desmoplastic small round cell tumour of childhood

Teatment Schedule

Following first laparotomy:

	Vincristine	Adriamycin	Cyclophosphamide	Etoposide	Ifosfamide
Course 1	2mg	30mg/m^2	600mg/m^2		
Course 2	2mg	30mg/m^2	600mg/m^2		
Course 3	0.4mg x 3 days	20mg/m^2 x 3 days	30mg/kg x 2 days		
Course 4				100mg/m^2 x 4 days	1g/m^2 x 4 days
Course 5	0.4mg x 3 days	25mg/m^2 x 3 days	50mg/kg x 2 days		
Course 6				100mg/m^2 x 4 days	1g/m^2 x 4 days
Course 7	0.4mg x 3 days	25mg/m^2 x 3 days	50mg/kg x 2 days		

Following 'Second-look' laparotomy:

	Vincristine	Adriamycin	Cyclophos
Course 8	0.4mg x 3 days	25mg/m^2 x 3 days	60mg/kg x 2 days
Course 9	0.4mg x 3 days	25mg/m^2 x 3 days	70mg/kg x 2 days

Followed by Radiotherapy (total abdominal) 3000 cGy (shielding of liver and kidneys at 1500cGy)

of childhood is made on the basis of its 'divergent differentiation' expressing markers of epithelium, nerve and muscle. The expression of α-fetoprotein, as in our case, is unusual. In addition, the nesting pattern of growth with surrounding intense desmoplastic reaction is characteristic of this variant of SRCT. Clinically it presents as a large abdominal tumour of the visceral peritoneum with multiple similar small peritoneal lesions[2]. These tumours are clinically and immunohistologically distinct from other SRCT such as Ewing's sarcoma, lymphoma, rhabdomyosarcoma, and primitive neuroectodermal tumours. Recently, a chromosomal translocation, t(11,22)(p13;q12), has been found to be associated with this tumour, and is responsible for the formation of a fusion gene between the EWS oncogene (located at 22q12) and the WT1 tumour suppressor gene (located at 11p13) which produces chimaeric messanger RNA[3]. This is different from the typical translocation seen in Ewing's sarcoma – t(11,22)(q24,q12) – which involves the same breakpoint on chromosome 22, but a different breakpoint on chromosome 11. Unfortunately we were not able to perform cytogenetic analysis on our case.

The treatment of this tumour is difficult, and the prognosis very poor. Complete surgical removal is recommended, but can rarely be achieved due to extensive spread within the peritoneal cavity. The tumour has been reported to be quite

chemo-resistant, but good partial responses have been reported[4]. Unfortunately, CT scanning may underestimate the intra-abdominal disease because small deposits may be missed, and therefore 'second-look laparotomy' is recommended[5]. Total abdominal radiotherapy and even bone marrow transplantation have been used as final consolidation, but the effectiveness of these regimes is not known[4]. Despite all these modalities of therapy, the majority of cases have already died by the time they are reported (e.g. 15 out of 19 in the series of Gerard *et al*[2], with a median survival of 15 months). Our patient showed disappearance of enlarged lymph nodes and a liver nodule on CT scan after VAC and etoposide/ifosfamide chemotherapy, implying a degree of responsiveness. However, nodules of residual disease were still found at second-look laparotomy. He underwent two more cycles of VAC and total abdominal radiotherapy, but without a further laparotomy we had no way of assessing his response to this. The dose of abdominal radiotherapy may have been too low to control such macroscopic disease. His response was short-lived, and he died of progressive disease. Further studies are required to delineate the best form of therapy for this highly aggressive tumour.

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