



British Sarcoma Group Conference 2009

ABSTRACTS – POSTERS

9005

Desmoid Tumours: A case series of 22 patients treated in Glasgow

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Introduction

Desmoid tumours are benign lesions derived from musculoaponeurotic structures. They are rare tumours accounting for 0.03% of all neoplasms with an estimated incidence of 2-4 per million per year. Women of child-bearing age are most commonly affected as well as an association with familial adenomatous polyposis. Whilst histologically benign, they can pose a significant surgical challenge with high rates of local recurrence.

We present the experience of a single surgeon specialising in the management of soft-tissue tumours.

Methods

We conducted a retrospective analysis of the West of Scotland Sarcoma Group soft-tissue tumour database. 22 patients with a histological diagnosis of fibromatosis were treated over a 13-year period between 1995 and 2008.

Results

The median age at presentation was 34 years. There was a marked female preponderance with a male: female ratio of 5: 17. 4 patients had fibromatosis in association with FAP and 3 presentations were related to recent pregnancy. The most commonly affected tumour site was the anterior abdominal wall (14) followed by the abdominal cavity (6), lower limb (2) and back (2). 2 patients presented with synchronous abdominal wall and intra-abdominal tumours. Of the superficial tumours 15 were excised with curative intent using mesh reconstruction where required, 2 were debulked, and 1 tumour was simply observed in a patient with learning difficulties. Of the 6 intra-abdominal tumours 5 arose from either small or large bowel mesentery. Of these 4 were resected with the involved intestinal segment and 1 is under observation and asymptomatic at 3 years follow up. The 1 pelvic tumour underwent local resection.

5 patients experienced minor wound morbidity but no major postoperative complications were observed. Tumour was evident microscopically at resection margins in 19 of the resected specimens. 4 patients recurred at a median of 22 months, 2 superficial and 2 intra-abdominal tumours. All have undergone repeat resection. At a median follow up of 39 (10-73.5) months one patient with Gardner's syndrome has died from metastatic gastric cancer.

Conclusion

Our data represents a single surgeon's experience managing a rare and challenging tumour. Negative resection margins can be difficult to achieve due to the anatomical position of the tumour and impalpable disease. Despite positive margins prognosis in this series of patients was good. Although recurrence rates can be high our data suggests that a 'wait and see' policy is safe as all recurrent disease was resected. There has been no mortality due to fibromatosis within the follow-up period.