

A REVIEW OF 10 YEARS OF SURGERY FOR FAMILIAL ADENOMATOUS POLYPOSIS ASSOCIATED DESMOIDS

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INTRODUCTION

- Rare <0.1% tumours
- Incidence 2-4/million/year¹
- RR desmoids in FAP
850 general population²
- 10-25% lifetime risk
- With duodenal disease most important cause of mortality in patients who have had a colectomy³



1 Reitamo et al 1982

2 Gurburz et al 1994

3 Arvantis et al 1990

INTRODUCTION

- Treatment difficult
- Poor quality studies
- Surgery recommended as treatment for abdominal wall and extra-abdominal desmoids
- Surgery for intra-abdominal desmoids generally avoided

INTRODUCTION

- 8/22 deaths post –op
- 6/14 survivors TPN
- 3 required >10units blood perioperatively¹
- obstructive complications, highly symptomatic



AIMS

- Reassess outcomes of surgery for desmoids in FAP over a 10 year period
- Do we need to revise our recommendations regarding the role of surgery in desmoid disease?

METHODS

- Surgery from 1994-2004 identified by search St Mark's Polyposis Registry database
- Registry records, hospital records and pathology reports examined

RESULTS

- 23 patients were identified
- 2 notes unavailable, insufficient data
- 1 patient has had FAP excluded since this review

RESULTS

- 14 female, 7 male
- Mean age at diagnosis 32 years (18-54)
- 31 desmoid tumours removed from 21 patients (mean 1.5, 1-6)
- 17 intra-abdominal (54.8%), 10 abdominal wall (32.3%), 4 extra-abdominal (12.9%)

RESULTS

- 15/21 colectomy prior to diagnosis
 - 2 colons still intact
 - 3 desmoid diagnosed before surgery
 - 1 incidental finding at colectomy
- Gap colectomy and desmoid diagnosis 5 years (1-19)

RESULTS

- Pouch (n=5), 2.2 years (1-4)
- IRA (n=9), 7.3 years (1-19)
- PPC (n=1), 2 years

RESULTS

- Pre-operative NSAID/ORM 19/21 (90%)
- Pre-operative chemotherapy 6/21 (28.5%)
- Post-operative NSAID/ORM 16/21 (76.2%)

RESULTS

- Indication for surgery intra-abdominal:
 - Pain 3
 - Fistulation to bowel 2
 - SB Obstruction 5
 - Failed medical treatment 2
 - Desmoid perforation 2
 - Incidental at GI surgery 3

RESULTS

- Operative mortality 0/31 (0%)
- Long term TPN 1/31 (3.2%)
 - 1/17 (5.9%) intra-abdominal desmoids
- SB resection info in 16/17 operations
 - Mean 30.4cm (0-200cm)
- Inoperable and bypassed 2/17
 - Emergency, perforation and peritonitis n=1
 - Elective, SBO

RESULTS

- Length of stay 12 days (1-90)
- Blood transfused 3 units (0-11)
- Complications 3/31 (9.6%; 2 abdo wall, 1 intra-abdo)
 - wound infection
 - return to theatre, post-op bleeding
 - re-admitted post-op with fever and vomiting

Results

- 20/31 surgical clearance clinically complete
- 8/20 fully excised recurred
 - 3 of 17 (18%) intra-abdominal
 - 1 of 4 (25%) extra-abdominal
 - 4 of 10 (40%) abdominal wall
- Follow up 5 years (0.6-10 years)

CONCLUSIONS

- 1) Surgery for intra-abdominal desmoids in carefully selected patients, is less hazardous than previously reported
- 2) Surgery for abdominal wall and extra-abdominal tumours is safe
- 3) Recurrence postoperatively remains a major problem

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