Medi-Scope Issue No. 11 Nov., 87

Malignant Melanoma of the Gastrointestinal Tract

DR. MARK BUGEJA MD
HOUSE PHYSICIAN - SURGEON

Malignant melanoma is a tumour which arises from melanocytes and usually proves to be rapidly fatal. It can occur at any site of the body where melanocytes exist including the choroid and ciliary body of the eyes, the skin and the ano-rectal region of the bowel. Ano-rectal malignant melanoma was first documented in 1857 by Moore. Over 400 such cases have been reported in the medical literature, up to 1984.

Incidence

3-15% of all tumours arising in the anal canal. 1-3% of all malignant melanomas develop at this site. Ranks behind only the skin and the eyes as source of primary malignant melanoma. In general melanomas effect females more often than males (3:1); maximal incidence occurs in the sixth decade; average age at diagnosis of primary lesion is about 42 years; average age at diagnosis of GIT metastases is about 46.5 years.

Pathology

Melanoma has an unusual predilection to GIT metastases. It is responsible for 50-70% of metastatic lesions to the small intestine (small intestine>colon> stomach). It spreads haematogenously and disseminates widely involving several organ systems. Visceral metastases (to liver, lung, bone) is associated with a very poor prognosis with life expectancy (1 year from the time of diagnosis). Malignant melanoma involving the GIT, with no evidence of a primary growth elsewhere in the body may be a primary but not necessarily so.

Aetiology

Exposure to U.V. radiation is thought to be responsible for cutaneous melanoma.

Clinical Features of GIT Involvement

Intestinal obstruction (cramping pain, vomiting); altered bowel habits; GIT bleeding (not massive, or chronic with + F.O.B's); anaemia; abdominal pain (intractable or non-specific); tenesmus; mass visible externally at anal margin or palpable during rectal examination; anorexia; nausea; vomiting; weight loss; pruritus; dysphagia; patients may present with a primary skin melanoma or metastatic enlargement of inguinal lymph nodes; unusual features include symptoms related to acute perforation of the bowel or malabsorption. A not uncommon complication is intussusception.

The time from initial diagnosis to GIT symptoms in GIT secondary melanomas has been found, in one study, to vary from 6 months to 90 months (mean approx. 43.5 months).

Differential Diagnosis

Malignant melanoma involving the anal canal may be mistaken for haemorrhoids or benign polyps. Hence, any tissue removed surgically from the anal region should always be sent for histological scrutiny.

Diagnostic Methods

Barium meal (for upper GIT masses)
Barium enema (for caecal or colonic masses)
Gastroduodenoscopy + biopsy
Proctosigmoidoscopy + biopsy
Abdominal Ultrasonography
Computerised Axial Tomographic Scan

These investigatory methods may reveal the melanomas as polypoid or ulcerating masses or intramucosal nodules in the case of gastric and colonic lesions. Radiographically ulceration of a gastric lesion gives rise to the *bull's eye* or *target* appearance. Small intestinal melanomas are polypoid in>50% of instances. These may act as leading points for intussusception. Colon may appear to be compressed by extrinsic masses. Endoscopy is helpful only if the lesion/s are accessible and particularly if they are pigmented; biopsies are mandatory.

Treatment

The treatment of choice is surgical. Adjuvant methods include chemotherapy (e.g. Dacarbazine, cyclophosphamide and radiotherapy, though experience of the latter has shown that no benefit is derived. In some places trials are being carried out using Thymostimulin, monoclonal antibody-directed effector cells and Human Fibroblast Interferon.

Methods

*Bowel resection (including Abdomino-perineal resection [or otherwise known as Combined Synchronous Excision]) - resection may be complete or partial

*Intestinal by-pass (shunting) procedures

*Combined resection and by-pass

*Some surgeons have also attempted bilateral inguinal and pelvic lymph node dissection when these were found to be enlarged due to metastases.

Indications for Operative Treatment

*Intestinal obstruction

*Intestinal bleeding

*Abdominal or rectal mass

*Intractable pain

*Anal melanoma

Aims

It is the general consensus that palliative operations in patients with GIT symptoms is indicated to relieve symptoms, to improve the quality of life and thereby perhaps helping to extend the duration of life. The operations are not intended to be aggressive i.e. an attempt made to remove as much of the tumour as is possible, since such procedures do not increase the life expectancy but, on the contrary, increase seriously both the morbidity and the immediate post-operative mortality. Without surgical palliation, the prognosis of melanoma patients with visceral metastases is very poor.

Results

No immediate post-operative deaths were recorded following conservative surgical interventions. Wound infections and complications associated with intestinal anastomoses were absent.

Some studies have shown no significant increase in the survival time and results were very grim (Pyper and Parks). Other investigators have shown significant benefit was derived from palliative surgery (Jorge et al; Freedman; Taylor et al; Reintgen et al). Many patients have been relieved of distressing GIT symptoms.

Survival time post-operatively varied from 1 to 47 months in most cases and up to 10 years in some even in spite of the presence of metastases (Freedman - vide infra). Overall, however, the prognosis remains poor irrespective of treatment.

Conclusions

1. Improvement in survival can be expected as greater public and professional awareness of the condition leads to earlier diagnosis and treatment.

2. Radiological methods of investigation are useful and reliable. Endoscopy is an excellent procedure to confirm the diagnosis in accessible regions. CAT scanning is helpful in identifying extensive hepatic, mesenteric and retroperitoneal involvement.

3. Spontaneous regression of original lesions is thought to occur and may explain the lack of primary lesions in some patients with metastatic melanoma.

4. Anorectal melanomas are anal primaries even if anus appears to be uninvolved. Abdomino-perineal resection is indicated and is curable in early cases.

5. Diagnosis is usually possible only after histological examination of involved tissue. All tissues excised from the anorectal region including haemorrhoids should be submitted for histologic assessment.

6. It is well recognised that tumour thickness is the best single prognostic indicator for melanoma. Tumours<2 cm in thickness have a better prognosis; exceptions have been documented. Bone, liver, lung and lymph node metastases appear to be associated with a very poor prognosis.

7. GIT surgery offers good relief to most patients with GIT symptoms, even in those with extra-GIT metastases.

8. Surgery should be limited to sites causing symptoms. Incidental lesions discovered during the operation should not be excised. Whether lymphadenectomy should be performed is still disputable; it is not done routinely in some centres and no longer practised in most.

9. No constant factor is apparent and to which increase in survival can be attributed.

10. Although associated with a poor prognosis, it is worth treating malignant melanomas of the anorectal region aggressively as there may be some unexpected good results.

References

Freedman: *Melanoma of the Anorectal Region - 2 Cases of Prolonged Survival* Brit.J.Surg. 1984 Feb: 71(2):164.

Jorge et al: *Symptomatic Malignant Melanoma of the Gastrointestinal Tract. Operative Treatment and Survival* An. Surg. 1984 Mar: 199(3):328.

Pezim and Nicholls: *Survival after High/Low Ligation of Inferior Mesenteric Artery during Curative Surgery for Rectal Cancer* Am. Surg. 1984 Dec: 200(6).

Pyper and Parks: *Melanoma of the Anal Canal* Brit.J.Surg. 1984 Sep: 71(9): 671.

Reintgen et al: *Radiologic, Endoscopic and Surgical Considerations of Melanoma Metastatic to Gastrointestinal Tract* Surg. 1984 Jun: 95(6): 635.

Seigal et al: *Surgical Treatment of Anorectal Melanomas* Am.J.Surg. 1983 Sep: 146(3): 336.

Cases Presented by Freedman (Brit. J. Surg. Feb: (1984) 71 (2): 164)

Case 1. A 31-year old male presented with rectal bleeding and diarrhoea of one year duration. On digital and proctoscopic examination, a 2 cm polypoidal lesion in anal canal was found. Biopsy was taken and resulted as Malignant Melanoma. Treatment: pre-operative external tele-cobalt radiotherapy: a single dose of 600 rads daily for 3 days (total=1800 rads): A.P. resection of rectum was performed. Liver was normal. Specimen sent for histology; post-operative vinblastine chemotherapy. Histology report: Poorly differentiated epithelioid type malignant melanoma)13 mm in depth. Pedical free of tumour. One mesenteric lymph node examined: partially replaced by tumour. Follow-up: 12 years later the patient was well and clinically free of metastasis.

Case 2. A 71-year old female patient presented with rectal bleeding and tenesmus of 6 months duration. On digital and proctoscopic examination a 1 cm roughened area was discovered posteriorly in the anal canal. Biopsies taken showed features of a poorly differentiated squamous cell carcinoma. Treatment consisted of a laparotomy with A.P. resection of rectum. Liver contained 3 black nodules (not biopsied). Histology revealed malignant melanoma of epithelioid type. At anorectal junction, tumour extended into submucosa; dermis only in anal skin. One mesenteric lymph node was examined: completely replaced by poorly differentiated tumour. Follow-up: 10 years later the patient remained well and there was no clinical evidence of recurrence.

Presentation of Case at St. Luke's Hospital, Malta

A 57-year old woman, presented in February 1985 with a 4 month history of constipation. The patient used to defecate once every 3 to 4 days. The stools were loose and frank (bright red) blood was passed but no mucus. She also experienced severe tenesmus. She did not suffer from any abdominal pain, nor nausea and vomiting; she did, however, complain of slight, non-specific dyspeptic symptoms. Appetite was always good; no significant weight change was noticed; there were no symptoms related to cardio-respiratory, genito-urinary and nervous systems. She was concurrently suffering from Diabetes mellitus.

Past History: R. Facial Palsy (several years ago): cholecystectomy (+appendicectomy) 9 months previously.

Social History: non-smoker, teetotaller, married and had 9 children.

On Examination: middle-aged, co-operative, healthy looking obese lady; not in pain; R. Facial palsy; afebrile; no clinical signs could be elicited during abdominal examination; Digital, rectal examination revealed a mass in the left wall of the rectum; inguinal lymph glands were not palpable.

On auscultation of the chest, rhonchi were perceived over the base of the left lung. Other systems were clinically normal.

Proctosigmoidoscopy: 2 to 3 cm from the anal verge, a cauliflower-like growth 2 to 3 cm in diameter situated on the left wall of the rectum was visualised. The rest of the rectum appeared normal. The growth was biopsied for histological evaluation.

Provisional diagnosis: Carcinoma of the rectum.

Biopsy Report: Malignant Melanoma.

Treatment: 1. Combined Synchronous Excision of the Rectum (with a terminal, permanent colostomy)
2. Chemotherapy (Dacarbazine)

Pathological scrutiny of the tissue removed at operation confirmed previous histological report of biopsy - Malignant Melanoma - with lymph node metastasis.

The patient died in October 1985 from hepatic failure and liver metastases.