

Primary Isolated Extramedullary Plasmacytoma of the Colon

A Case Report

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SUMMARY

A tenth case of extramedullary plasmacytoma (EMP) of the colon is reported. The term 'primary isolated extramedullary plasmacytoma' is preferred to those terms previously used. A broad classification of plasmacytoma of the gastro-intestinal tract is suggested and a management policy, based on this classification, is outlined.

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Plasmacytoma of the colon or rectum is an uncommon lesion. Only 9 cases have previously been reported in the English literature. Of these, only 4, by virtue of adequate documentation, can be regarded as primary isolated plasmacytomas of the large bowel.¹⁻³ This implies that there is no evidence of bone marrow plasmacytosis, Bence Jones proteinuria, or radiological changes in bone.

A tenth case of extramedullary plasmacytoma (EMP) arising from the large bowel is presented. It is the fifth case of primary isolated EMP reported as having occurred at this site. Of the previously reported colonic plasmacytomas 5, by virtue of specific positive findings of generalized plasma cell dyscrasia or the absence of documented exclusion of such manifestation, cannot truly be classified as primary and isolated. The patient reported by Hampton and Gandy,⁴ in whom the rectum was involved, had undergone resection of a previous plasmacytoma of the stomach, and the lesion cannot therefore be considered primary.

CASE REPORT

A 61-year-old White man was admitted to the surgical ward for Billroth I gastrectomy for a gastric ulcer, diagnosis of which was confirmed by barium meal examination and fibre-optic gastroscopy. Results of investigations on admission were as follows: haemoglobin 12.1 g/100 ml, white cell count 9 400/ μ l and ESR 75 mm/1st h (Westergren). Electrolyte and acid base status, and liver and renal function were normal. The chest radiograph was normal.

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and the ECG showed mild left ventricular strain. A positive occult blood test on rectal examination was ascribed to his gastric ulcer.

On 3 March 1976, the abdomen was opened via an upper midline incision. Routine examination of the abdominal viscera disclosed a mass in the distal sigmoid colon. Since the colon had not been prepared, no resection was attempted. An enlarged lymph node of the inferior mesenteric group was removed for histological examination. This was subsequently reported as having shown 'reactive changes only'. A gastrectomy was not performed.

After an uneventful recovery of the patient, a barium enema examination (Fig. 1) supported the suspicion of a malignant lesion of the sigmoid colon, beyond the reach of a rigid sigmoidoscope. Colonoscopy was not available to us.

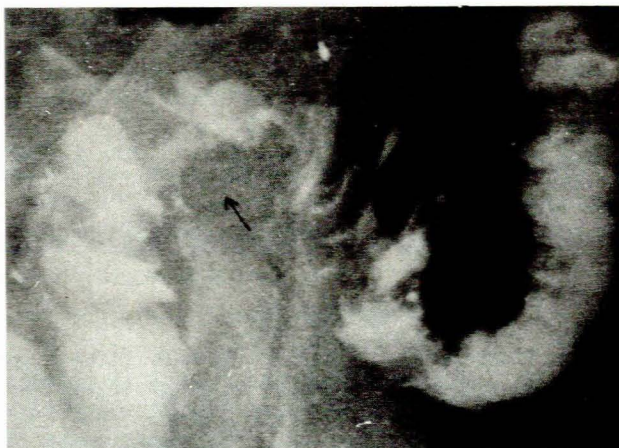


Fig. 1. Radiograph of barium enema examination, showing the filling defect (arrowed) in the distal sigmoid colon.

After adequate pharmacological and mechanical bowel preparation, the patient underwent a sigmoid colectomy on 7 April, with primary end-to-end anastomosis. Post-operative recovery was uncomplicated.

Histology of the lesion (Fig. 2) was reported on as follows:

'Histology is that of a plasmacytoma, with sheets of plasma cells, some of which are binucleated. Mitotic figures are infrequent. Calcification of the tumour and submucosa is present, and there are iron pigment deposits in the former. The tumour is well circumscribed, involving the mucosa and submucosa but also extending into the muscularis propria. Superficial ulceration and secondary

acute inflammatory changes are present. The tumour is surrounded by focal areas of lymphocytic infiltration and the regional lymph nodes show non-specific reaction. Total excision is presumed, as the limits of the specimen are through normal tissue. Scattered diverticula are present in the colon.'

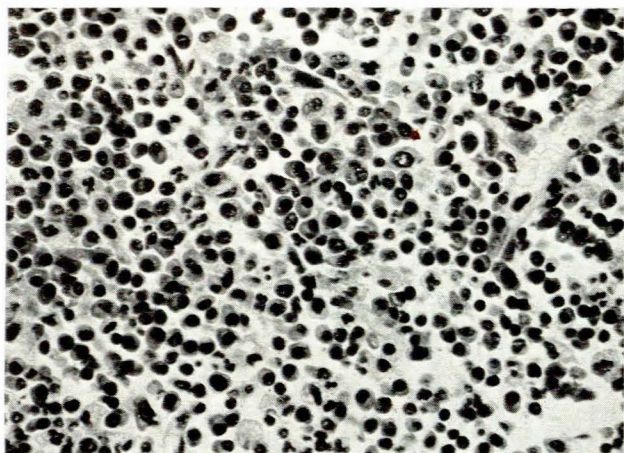


Fig. 2. Section of tumour showing sheets of plasma cells, a few larger plasmablasts, and a paucity of mitotic figures (H and E $\times 400$).

In view of the diagnosis, further investigations were performed. Repeat screens of liver and renal function were normal. The serum uric acid level was within normal limits. There was no proteinuria, no Bence Jones protein, and the serum albumin level was normal, with a normal globulin pattern on electrophoresis. Bone marrow aspirate and trephine biopsy showed no evidence of myeloma. Radiological skeletal survey was normal.

Repeat gastroscopy on 15 April revealed that the gastric ulcer had healed.

DISCUSSION

Dolin and Dewar⁸ have classified neoplastic plasma cell proliferation as (i) multiple myeloma; (ii) solitary myeloma of bone; (iii) plasma cell leukaemia and (iv) EMP. They state that the EMPs do not exhibit the manifestations of disturbed protein metabolism. Demonstration of abnormal proteins (paraproteins) would therefore preclude the diagnosis of primary isolated EMP. Others,^{1,6} however, be-

lieve that production of an M protein by the cells of such a tumour is compatible with the diagnosis, provided that disappearance of the monoclonal peak follows complete resection.

A further distinguishing feature is lymph node involvement. This is common in primary isolated EMP, but is seldom found in other foci of plasma cell neoplasia.³ In a comprehensive review, Wiltshaw⁶ has stressed the importance of regarding primary isolated EMP as a pathological entity separate from other plasma cell dyscrasias. This distinction has a bearing on the modalities of treatment and on prognosis.

Approximately 10% of all EMPs are found in the gastro-intestinal tract,^{3,7,9} but fewer than half of these may be regarded as primary and isolated.⁶ Extramedullary plasmacytoma of the bowel has been described as part of multiple myelomatosis.¹⁰ The majority of primary isolated EMPs show evidence of malignancy, but subsequent development of generalized disease is uncommon,¹ and should probably cast doubt upon the original diagnosis.

We feel that all primary isolated EMPs should be considered malignant¹¹ and treated accordingly. Most authors recognize the role of local resection and irradiation, but chemotherapy is of dubious value.^{2,5} It should be considered, however, when there are distant metastases.⁶

EMP, although solitary, but part of a generalized plasma cell dyscrasia, should be treated locally on its merits, in addition to conventional aggressive treatment for myelomatosis.

A rational systematized approach to the management of EMP includes the following investigations: full blood count, peripheral smear, and estimation of erythrocyte sedimentation rate; blood urea, serum creatinine, and creatinine clearance; liver function studies, and serum protein electrophoresis; immunodiffusion and immunoelectrophoresis; urine analysis for Bence Jones protein; bilateral trephine bone marrow biopsy (repeated biopsies at annual follow-up unless otherwise indicated) and radiological and isotopic skeletal survey.

These parameters are assessed at the time of diagnosis of EMP and at 6-monthly intervals after treatment, to exclude synchronous or metachronous plasma cell dyscrasia.

Treatment of gastro-intestinal EMP is based on its classification into either primary isolated EMP (and its staging) or EMP associated with synchronous or metachronous generalized disease (Table I).

TABLE I. CLASSIFICATION, STAGING AND TREATMENT OF GASTRO-INTESTINAL EXTRAMEDULLARY PLASMACYTOMA

Classification	Staging	Treatment
Group A	Stage I (localized disease)	Resection
	Stage II (lymph nodes or residual tumour)	Resection and radiotherapy*
	Stage III (distant disease)	Resection, radiotherapy and chemotherapy†
Group B	Synchronous generalized disease	Resection if indicated, radiotherapy, and chemotherapy
Group C	Metachronous generalized disease	Chemotherapy

* Radiotherapy: local field and regional nodal irradiation.

† Chemotherapy: full therapy as for multiple myeloma.

Group A constitutes primary isolated EMP. This implies no stigmata of plasma cell dyscrasia, although evidence of an M component which disappears after complete resection does not preclude this diagnosis. Infiltration of adjacent tissue by the tumour, which is nevertheless resectable, still leaves the tumour in stage I. Histological evidence of incomplete resection warrants reoperation, if feasible. If not, the tumour should be restaged as stage II. Distant metastases automatically indicate stage III disease.

Resection of a gastro-intestinal EMP associated with synchronous disease (group B) should be performed if indicated, e.g. by intestinal obstruction or haemorrhage.

The appearance of metachronous generalized disease (group C) is unusual, but may occur.⁴ The original diagnosis of primary isolated EMP should be reviewed. If the

only evidence of possible metachronous disease is the re-appearance of a monoclonal peak, local recurrence of the tumour must be excluded before chemotherapy is considered.

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Infantile Cortical Hyperostosis (Caffey's Disease)

A Case Report

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SUMMARY

An infant in hospital unexpectedly developed infantile cortical hyperostosis (Caffey's disease) while undergoing treatment for an unrelated illness. The presentation of the disease was classic and there was marked thrombocytosis. The aetiological possibilities are discussed.

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Caffey's disease or infantile cortical hyperostosis was described by Caffey and Silverman in 1945. The disease is characterized by subperiosteal new-bone formation involving multiple bones and associated with tenderness and swelling of the overlying soft tissues. It is a rare disease with an unknown aetiology. An unusual feature of the case we wish to present is that it developed while the patient was in hospital undergoing treatment for a

different illness. Furthermore, it is apparent from the circumstances surrounding the patient's illness that an infective aetiology cannot be dismissed. The striking thrombocytosis that may occur during the course of the illness is also documented.

CASE REPORT

The patient was a White female infant who was born at home on 9 March 1975, following a normal vertex delivery. Her birth weight was 2.46 kg. No maternal illness was recorded during pregnancy and no problems arose during the neonatal period. At 3½ months of age the patient developed a urinary tract infection and an excretory urogram demonstrated an obstruction of the left pelvi-ureteric junction. She was referred to our hospital for surgical correction of this anomaly. On admission her weight was recorded as 4.7 kg (3rd Boston percentile) and her length and head circumference measurements were between the 50th and 75th percentiles respectively. There were no other siblings in the family.

Examination revealed generalized hypotonia with some delay in motor development. The rest of the examination revealed no abnormality. An X-ray film of the chest taken at this stage was normal (Fig. 1). Before transfer to our hospital the haemoglobin level was 8.9 g/100 ml

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