The Ulster Medical Journal, Volume 55, No. 2, pp. 187 – 189, October 1986.

Case report

Primary malignant lymphoma of the appendix

R J Stewart, M Mirakhur

Accepted 1 August 1986.

Primary malignant lymphoma of the appendix is a rare tumour. We report a case and review the literature discussing the management and prognosis.

CASE HISTORY

A 33-year-old female presented with an acute onset of lower abdominal pain of sixteen hours' duration. She gave a three-month history of general malaise but was otherwise asymptomatic. On examination she was pyrexial, temperature 37.5°C with lower abdominal tenderness and rebound. The haemoglobin was 12.9 g/dl and the white cell count 16 × 10^3 µl with a normal differential. A gynaecological opinion was requested, vaginal examination revealing a fullness in the right side of the pelvis, which on ultrasound scan was compatible with an ovarian cyst. Laparotomy through a Pfannensteil incision revealed a 7 × 4 cm tumour in the body of the appendix which was perforated. The caecum was normal. In view of limited access, appendicectomy plus resection of adjacent caecum was performed, following which the patient made an uneventful recovery. Postoperatively her leucocytosis resolved and her white cell count to date remains within normal limits.

Macroscopically the resected specimen showed the appendix to be distended by greyish-white tumour which was infiltrating along the wall into the caecum. Histologically the lesion was a diffusely infiltrating lymphoma replacing the normal lymphoid tissue of the appendix, the limits of resection being free of tumour. On immunoperoxidase staining, the lesion was confirmed to be a B-cell lymphoma of the centrocytic/centroblastic type.

Subsequent screening, with a small bowel series, bone marrow examination and CT scan, did not reveal the presence of further tumour. A chemotherapy regimen consisting of vincristine, cyclophosphamide and prednisolone was commenced three weeks after surgery, and eighteen months later the patient remains free from relapse.

DISCUSSION

Malignant lymphomas comprise one to four per cent of malignant neoplasms of the gastrointestinal tract.[1] Primary involvement of the bowel occurs in five per cent of all lymphomas[2] of which 33 – 63 per cent affect the stomach, 25 – 60

Departments of Surgery and Pathology, The Queen's University of Belfast.
R J Stewart, FRCS, Surgical Registrar.
M Mirakhur, MD, MRCPath, Senior Medical Officer.

Correspondence to: Mr R J Stewart, Department of Surgery, The Queen's University of Belfast, Institute of Clinical Science, Grosvenor Road, Belfast BT12 6BJ.

per cent the small intestine and 3 – 30 per cent the large intestine.[1-4] Sole involvement of the appendix is rare, Collins reporting 11 cases in 71,000 appendix specimens, an incidence of 0.015 per cent.[5] A review of the literature shows all tumours reported to be of the non-Hodgkins variety, presenting from 4 – 55 years (mean 18 years) and with a male predominance of 1.5:1.[1, 6-12]

The commonest presenting symptom was right iliac fossa pain recurring over a period of months, frequently associated with a palpable mass. A lesser number presented acutely with systemic features or 'appendicitis', our patient developing peritonitis secondary to tumour perforation. This complication explains her leucocytosis which rapidly resolved postoperatively.

Operative treatment may be by appendicectomy with or without resection of caecum or a right hemicolectomy. In the event of resection not being possible tumour limits should be outlined by metal clips to facilitate subsequent radiotherapy. At operation the abdomen should be thoroughly explored for involvement of regional and distant lymph nodes, hepatosplenomegaly and multicentric lesions within the gut.

Postoperative management may comprise radiotherapy, systemic chemotherapy or a combination depending on the histopathological features and stage of disease. Irradiation is a potent agent for achieving local control and is curative in over 50 per cent of patients with stage I and II non-Hodgkin's lymphoma.[13] With the identification of effective combination chemotherapy for advanced lymphoma, it is not surprising that these drugs have also been utilised for patients with stage I and II disease, with results as good and probably better than radiotherapy alone.[14] Combined modality therapy reports excellent results for stage I and II disease but shows no improvement in advanced disease to date.[15]

Details regarding survival of patients with a primary lymphoma of the appendix are scanty, with reports varying from one month to 28 years. In the absence of adequate staging and postoperative treatment, few conclusions may be drawn, but an early presentation, as with this patient, and adequate treatment should result in a prolonged survival.

We are grateful to Mr S T D McKelvey and Dr P Abram for permission to report this case.

REFERENCES

1. Loehr WJ, Mujahed Z, Zahn FD, Gray GF, Thorbjarnarson B. Primary lymphoma of the gastrointestinal tract: a review of 100 cases. *Ann Surg* 1969; **170**: 232-8.
2. Ike BW, Rosenbusch G. Gastrointestinal malignant lymphoma: roentgenographic features and pathologic and morphologic correlations. *Diagn Imaging* 1981; **50**: 66-80.
3. Kahn LB, Selzer G, Kaschula ROC. Primary gastrointestinal lymphoma. *Am J Dig Dis* 1972; **17**: 219-32.
4. Isaacson P, Wright DH, Judd MA, Mepham BL. Primary gastrointestinal lymphomas: a classification of 66 cases. *Cancer* 1979; **43**: 1805-9.
5. Collins DC. 71,000 human appendix specimens. A final report summarising 40 years' study. *Am J Proctol* 1963; **14**: 365-81.
6. Gilick DD, Soule EH. Primary malignant lymphoma of colon or appendix. *Arch Surg* 1966; **92**: 144-51.

7. Lewin KJ, Ranchod M, Dorfman RF. Lymphomas of the gastrointestinal tract. A study of 117 cases presenting with gastrointestinal disease. *Cancer* 1978; **42**: 693-707.

8. Knox G. Lymphosarcoma primary in the appendix. A study of 23 cases. *Arch Surg* 1945; **50**: 288-92.

9. Sin IC, Ling ET, Prentice RSA. Burkitt's lymphoma of the appendix: report of two cases. *Hum Pathol* 1980; **11**: 465-70.

10. Galloway WH, Owens EJ. Primary lymphosarcoma of appendix occurring in childhood. *Br Med J* 1949; **2**: 1387-8.

11. Schmutzer KJ, Bayar M, Zaki AE, Regan JF, Poletti JB. Tumors of the appendix. *Dis Colon Rectum* 1975; **18**: 324-31.

12. Clarke RG, Simonds JP. Primary lymphosarcoma of the appendix. *Cancer* 1951; **4**: 994-8.

13. Hoppe RT. The role of radiation therapy in the management of the non-Hodgkin's lymphomas. *Cancer* 1985; **55**: 2176-83.

14. Cabanillas F, Bodey GP, Freireich EJ. Management with chemotherapy only of stage I and II malignant lymphomas of aggressive histologic type. *Cancer* 1980; **46**: 2356-9.

15. Glatstein E, Donaldson SS, Rosenberg SA, Kaplan HS. Combined modality therapy in malignant lymphomas. *Cancer Treat Rep* 1977; **61**: 1199-1208.

BOOK REVIEW

Aids to the examination of the peripheral nervous system. (New ed.). (pp 61. Illustrated. £2.95). London: Baillière Tindall on behalf of the Guarantors of Brain, 1986.

This slim volume details the wiring diagram of the limbs and the methods of testing individual muscles with beautiful black and white illustrations. I have examined this new edition with interest. The previous edition has taken its place in my bag beside my ophthalmoscope and tendon hammer as an essential tool of the trade of a practising clinical neurologist. The new edition has retained the style of the old, being commendably brief and clear. It has made good one significant lack in previous editions, with the addition of a coloured diagram of the lumbosacral plexus.

There is something in this book for every student of neurology, no matter how old. At any price it is a bargain, but at its present price every student should have one. I hope the new publishers make it their business to ensure that there is a steady supply in the bookshops. Previous editions have gone out of print rapidly.

SAH