Primary Malignant Lymphoma of the Appendix

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ABSTRACT: A 70-year-old Japanese woman with primary malignant lymphoma in the appendix was treated. The diagnosis was established after surgery. Histologically, the tumor was malignant lymphoma, lymphocytic, well differentiated, according to the Rappaport's classification which is a good prognostic type of extranodal malignant lymphoma. The patient was treated by ileocecal resection and without radiochemotherapy. The 36-month follow-up revealed neither evidence of recurrence nor metastases.

KEY WORDS: malignant lymphoma, appendix vermiformis

INTRODUCTION

Although the gastrointestinal tract is the most common site of extranodal lymphoma, the appendix is rarely involved. The malignant lymphoma herein reported was confined to the appendix and its immediate mesenteric lymph nodes, as determined postoperatively. This case is of interest because of the advanced age of the patient. Adding the present case, 39 such cases reported in the literature are briefly reviewed.

CASE REPORT

A 70-year-old Japanese woman was admitted because of a painless, palpable tumor in the right lower quadrant of the abdomen. She had found it herself about ten days prior to admission. Loss of weight, abnormal bowel movements or other symptoms were nil. There was nothing relevant in her family or personal history.

Physical examination revealed a palpable, painless tumor, approximately 15 × 5 cm in size, in the right iliac fossa. Neither general lymphadenopathy nor hepatosplenomegaly was noted. Other physical examinations showed no abnormal findings.

On admission, roentgenologic examinations of the chest and abdomen revealed entirely normal conditions. Hematologic and serologic examinations were within normal limits, except for a positive CRP. Neither barium enema examination nor colonoscopy was performed. An ultrasound examination of the abdomen revealed a hypoechoic solid mass measuring 14 × 7 cm in the iliac fossa.

The tentative preoperative diagnosis was mucocele of the appendix. Following a right rectus incision, we found a huge sausage-shaped appendix. Three elastic, hard, mesenteric lymph nodes, measuring 3 × 2 cm, 2.5 × 1.5 cm, and 1 × 1 cm, respectively, were present in the ileocecal area. Ileocecal resection with mesenteric lymph node dissection was performed. Neither hepatosplenomegaly
nor swelling of distant lymph nodes were noted.

She was discharged and postoperative radiochemotherapy was not prescribed. Thirty-six months later she is well with no signs of recurrence.

Pathologic findings

The appendix was firm in consistency, measured 15 cm in length and 8 cm in diameter, with a smooth and partly nodular external surface (Fig. 1). The surface was whitish yellow with some purplish discoloration. When the specimen was opened along the longitudinal axis, the lumen of the appendix was filled with firm, yellowish, tumor tissue without covering mucosa (Fig. 2). The ileal end and cecum seemed to be free of tumor.

Histologically, the greatly enlarged appendix showed compact sheets of small lymphoid cells which involved the whole thickness of the wall. The normal architecture was lost. Most of the tumor cells had infiltrated diffusely. The nucleus of the cells was round and compact with inconspicuous nucleoli. Mitotic figures were rare (Fig. 3). Plasmacytoid differentiation was absent and intracytoplasmic immunoglobulins were not identified by the peroxidase-antiperoxidase (PAP) technique. In some areas, a vague nodular pattern usually found in small cell type of B cell lymphoma was recognized, as shown in Fig. 4. In these areas, the tumor cells were slightly

Fig. 1. Enlarged appendix with adjacent ileocecum and lymph nodes.

Fig. 2. The resected specimen opened along the longitudinal axis. The lumen of the appendix was filled with yellowish tumor tissue.

Fig. 3. Diffuse infiltration of small lymphoma cells (HE, ×500). No plasmacytoid differentiation.

Fig. 4. An area showing a vague nodular pattern (HE, ×37).