Primary Lymphoma of the Spleen

DALAĞIN PRIMER LENFOMASI

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SUMMARY

Involvement of the spleen is seen in a discrete number of patients with non-Hodgkin's Lymphoma (NHL). Splenic involvement can be pari of a diffuse dissemination of NHL in which the spleen is one of multiple involved organs or sites. Alternatively, non Hodgkin's lymphoma may also originate in the spleen and then spread to other sites. PSL does not spread in a predictable fashion. Thus, once the disease baccmes clinically evident, it may be difficult to appreciate the precise sequence of tumor progression.

Key Words: Non Hodgkin's lymphoma, spleen

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ÖZET

Dalağın tutulması Hodgkin dışı lenfomalı hastaların ayn bir bölümü olarak görülür. Hastalığın dalağa yayılması çok sayıda organ veya yerlere yayılmış olan Hodgkin dışı lenfomanın (NHL) dalağı diffuz olarak infiltre etmesidir. Diğer taraftan NHL dalaktan kaynaklanarak diğer taraflara yayılabilir. PSL'mn yayılma tarzı bilinmemektedir. Bundan dolayı klinik bulgular belirgin olduğunda tümör ilerlemesinin kesin sırasını belirlemek güç olabilir.

Anahtar Kelimeler Hodgkin dışt lenfoma, dalak

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Lenfomas, are primary malignancies of lymphatic tissues, originating from malign conversion of hystiocytic and lymphocytic cells. N H L is a unique group of malignant diseases, originating from immune system cells. Lenfomas were first described by Malpighi in 1766 (1). Hodgkin showed the gross anatomy of this disease in 1837, Stenberg in 1898 and Reed in 1902 described the disease antity, Oberling in 1928 and Roulet in 1930 reported the histologic description of the subgroups (1,2,3,4,5).

PRESENTATIN OF CASE

A 60 years old male, known to have loss of appetite, fatigue, weight loss and protrusion of the left hypochondrium, was brought to the hospital. On examination, a mass 15-16 cm. in diameter was palpable at the left upper quadrant of the abdomen. Hematologic studies showed no pathologic findings other than a high ESR (31 mm. in 1/2 Hr., 64 mm. in 1 Hr.). CT and abdominal ultrasonography showed an enlarged spleen measuring 15cm. at its longest diameter, Idler and paraaortic lymph nodes were not enlarged. Bone marrow aspiration was also normal.

Splenectomy was performed through a left subcostal incision (Picture 1). Exploration of the abdomen revealed that the Idler and para aortic lymph nodes were not involved. An incisional biopsy of the liver was performed. The histologic examination of the spleen was reported as consistent with "prolymhocytic lymphoma" (Picture 2), and the liver tissue was free of disease.

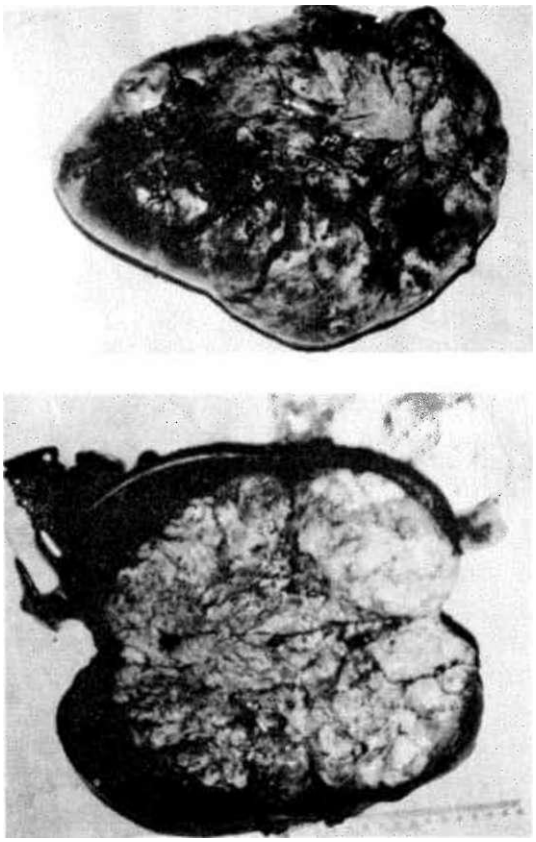

Figure 1. Spleen, removed by laparotomy.

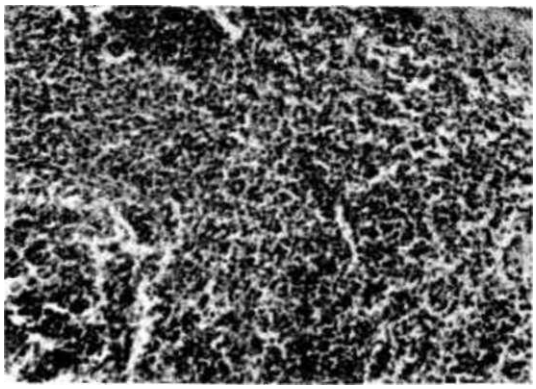

Figure 2. Histologic view of the spleen.

DISCUSSION

The etiology of NHL is not well known. NEL originating from immune system cells constitues a separate group of malignancies other than chronic lymphocytic leukemia, multipl myeloma and mycosis fungoides. Different studies were done to classify NHL and Brill in 1925, Symmers in 1927 have summed up NHL in two subgroups, nodular and follicular (7,8). Rappaport in 1966 made a new classification of NHL, which is accepted by most of the authors (9). Rappaport divides NHL to five subtypes. These are:

1. Lymphocytic differentiated lymphoma,
2. Lymphocytic indifferentiated lymphoma,
3. Lymphocytic-histiocytic lymphoma,
4. Hysticoytic lymphoma,
5. Indifferentiated cell (stem-cell) lymphoma.

The subtypes, other than indifferentiated lymphoma are also divided into two groups as nodular and follicular. NHL usually originates from lymph nodes, bone marrow, liver, digestive tract and skin. Involvement of the spleen is usually secondary and in rare instances primary. NHL originating from the spleen is known as "Primary Splenic Lymphoma". Spread the other organs occur from PSL. The route of spread to other organs are not known (10,11,12).

According to Ahmann PSL is classified in three groups (12).

1. Group I. Only the spleen involved
2. Group II. Spleen and hiler lymph nodes are involved.
3. Group III. Besides spleen and hiler lypmh nodes, other lypmh nodes, liver, bone marrow etc. are involved.

The significant finding of PSL is the enlargement of the spleen. But, significant peripheral lymphadenopathy is not present. It may spread to the regional lymph nodes, liver and bone marrow. Clinical findigns may be confusing because of Hypersplenism and pancytopenia (12,13,14,15). Patients with PSL, who undergo splenectomy because of pancytopenia or splenomegaly, resembles patients with NHL having spleen involvement (12). The existence of PSL can be understood, only after histologic studies of the spleen in patients with splenomegaly, who are known to have malignancies according to hematologic studies. The cilinical outcome of PSL is smoother than NHL involving the spleen (12).

Almost every patient with PSL, who are rarely found, have hematologic abnormalites even though it may be mild. Hematologic disorders are rare in

the studies of Long, Skarin and Hyatt, but more frequent in the studies of Ahman and Narong (13,14,15). Same authors report that hematologic abnormalites return to normal after splenectomy. Survival is longer in PSL compared to NHL. In Ahmann's report 31% of his cases survived for five years (13). According to Long and Aisenberg life expectancy is shorter (16). Narong's studies gives a longer survival for cases with spleen involvement (Group 1) compared to patients with hiler lymph and other organ involvements (Group II-I1I) (15,17,18).

Spread to the bone marrow decreases survival (12). The hematologic studies, abdominal ultrasonograpy and GT has significant place in diagnosis of the disease (19,20). Even though studies are undertaken with MRI recently, it has an insignificant place in diagnosis (21,22). Final diagnoses can be obtained by histologic examination (12). Treatment is primary surgical. Splenectomy is required for the diagnoses and removal of malignant mass and hypersplenism. After splenectomy radicating and combined chemotherapy should be used (6,12,23,24,25).

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