ACTA MEDICA IRANICA
Vol. XIV,1971,p.159-163

Lymphosarcoma Associated with Rheumatoid Arthritis and Facial Edema

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The relationship of autoimmune disease to lymphoproliferative neoplasma has been discussed by several authors (1,2,4,6). It appears that in immunological disorders the hyperactivity of lymphatic tissue leads to proliferation with occasional progression to lymphoma; conversely, in malignant lymphoma, the hyperplasia of immunological competent cells leads to autoimmune phenomena.

The case of a child with malignant lymphoma who developed migratory inflammatory arthritis is discussed. The object of the present paper is to document the coexistence of inflammatory polyarthritis and lymphosarcoma and to discuss analogous cases from the literature.

Case Report

D.F., a 14 year old boy, was admitted to the Pahlavi Medical Center on June 30, 1969, because of left cervical lymphadenopathy. Biopsy revealed malignant lymphoblastic lymphoma. He was treated with nitrogen mustard

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— Received for publication October 1971.

and discharged in good health on July 3, 1969.

He did well until the second admission on November 23, 1969 when he was suffering from pain in both elbows. Physical examination on admission revealed the cervical nodes to be 4 × 4 × 3 cm on the right side and 3 × 3 × 2 cm on the left. Both elbow joints were tender, hot, swollen and could not be extended. Heartbeat was normal. Two days after admission, the right knee became tender and swollen. The elbows were no longer tender or hot, but could not be extended well. Large lymph nodes were felt in the posterior areas of both elbow joints. Two days later the left wrist became tender and swollen. Five days later the right ankle became slightly swollen and tender. Pain in the other joints had disappeared, but the limitation of elbow joint movements persisted.

Three days later on December 9, 1969, the left ankle became somewhat tender, hot and slightly red and the patient developed severe edema on the face. Small amounts of fluid were detected in the right knee joint. The lymph nodes behind the elbows were 6 × 6 × 4 cm. Bone marrow test showed 90 % lymphoid infiltration, of which 40 % were lymphoblasts.

Laboratory Tests

Hemoglobin = 15.5 Gm/100 ml, white blood count = 7,600, eosinophil = 3 %, polymorphonuclear = 61 %, lymphocyte = 21 %, Monocyte = 12 %, lymphoblasts = 3 %; platelets and red blood cell were normal. Sedimentation rate was 48mm/hour.

Latex flocculation was negative. Protein electrophoresis: Total protein = 7.6 Gm %, albumin = 47.8 %, 1 globulin = 4.6 %, α 2 globulin = 12.4 %, β globulin = 13 %, γ globulin = 22.2 %. Immunoelectrophoresis showed very slight elevation of γ M. Total bilirubin, thymol turbidity and cephalin flocculation were normal. L.E. test was repeatedly negative. ASO titer was 250 Todd units. Chest x ray revealed enlargement of the left mediastinal nodes. Elbow joint x rays were negative.

After completion of the above studies therapy was began. He was last seen on October 3,1970, when the cervical nodes were quite large (12 x 14 x 8

cm on the right and 12 x 10 x 8 cm on the left). The spleen was palpable 2 cm below the left costal margin. The left wrist was somewhat swollen, hot and tender; joint x Ray was negative. Hemoglobin, 11.2 white blood cell = 24,000, platelets = 15,000, red blood cells = 4,700,000, peripheral blood showed 90 % of lymphoblasts.

Uric acid was 5.6 mg %, latex flocculation 1 +, sedimentation rate was 47 mm/1 hour.

Sensitized sheep red cell agglutination (Rose Waaler test) was 1/256 positive. Protein electrophoresis revealed albumin 53 %, α 1 globulin 3 %, 2 globulin 9 %, 3 globulin 11 %, γ globulin 24 %., Immunoelectrophoresis showed marked elevation of γ M (figure 1) which was evaluated at more than 130 mg % (normal 50-100 mg %).

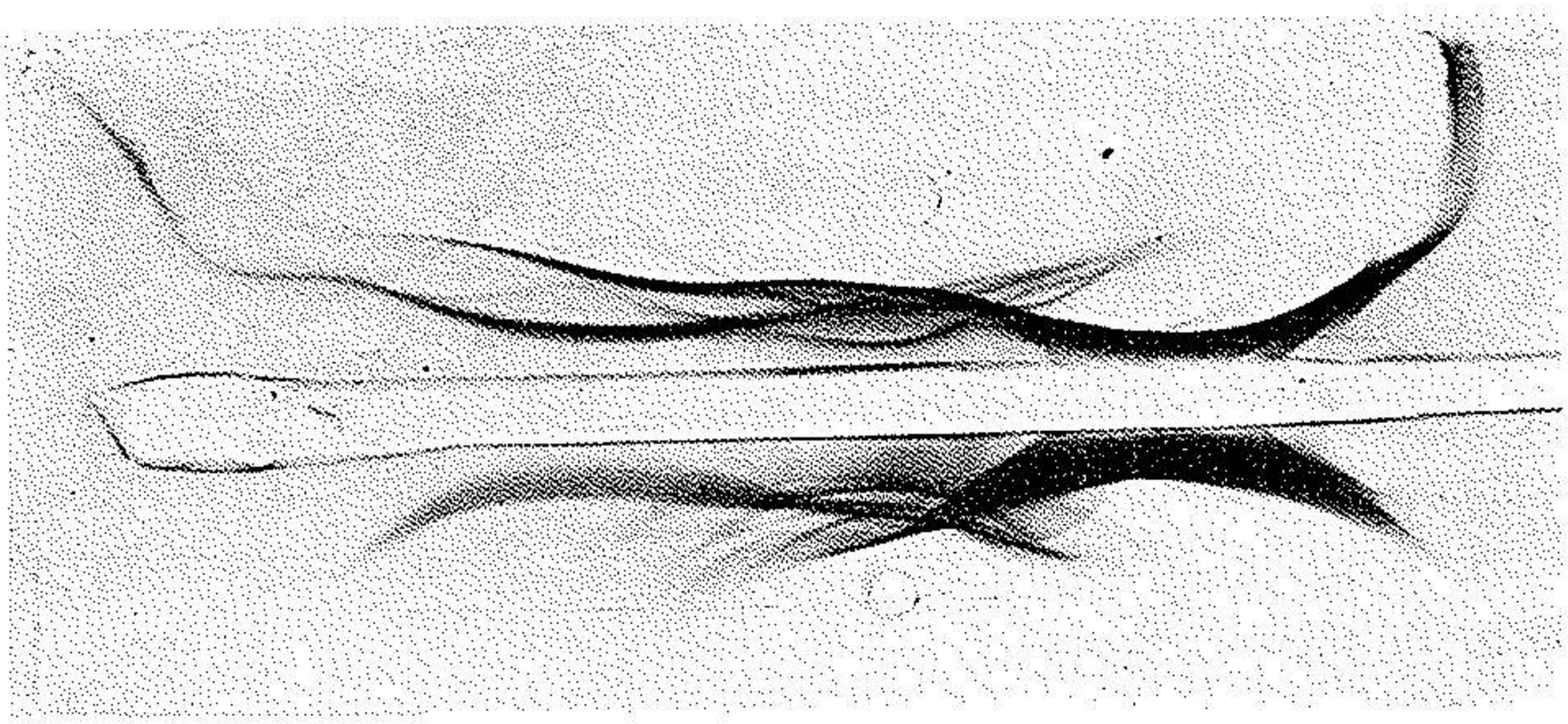

Fig. 1. Immunoelectrophoresis of sera protein
Upper well: serum D.F. 9.3. 1970
Lower well: normal human serum.

Discussion

Several authors have described the coexistence of immunological diseases of various kinds with malignancies of the lymphatic system (1,3,6,7,). Indeed it has been suggested that the two share a common etiology (1,7) even though this is disputed by some writers (4,8). In immunological disorders excessive activity of the lymphatic tissue may bring about cell proliferation and eventually frank lymphoma (1,7).

The cases reported in the literature (1-4,6,7) suggest that association of malignant disease of lymphatic tissue with joint symptoms is more common in adults than in children and that most frequently, malignant lymphoma occurs with lupus erythematosis or with rheumatoid arthritis.

The patient under discussion developed, hot, swollen, painful joints five months after the diagnosis of lymphoma was made. The clinical picture was suggestive of rheumatic fever. The sedimentation rate was quite high, the latex flocculation, immunoglobulins, L.E. preparation were repeatedly negative.

On the last visit there was a new development in the immunoelectrophoresis which revealed marked elevation of γM globulin; Rose-Waaler test was also highly positive. These findings made the diagnosis of rheumatoid arthritis more probable. When bone marrow tests were performed we were surprised to find marked infiltration of lymphoid cells. While extension of malignant lymphoma to distant areas is common and occurs in 67% of the cases (5) this type of presentation is exceedingly rare.

In addition to the joint manifestations, another unusual feature in this case was the marked facial edema, appearing a week after the onset of joint symptoms. This resembled the scleredema of Buschke which could be related to the malignant lymphoma. Analogous examples were not however, found in the literature.

Association of migratory inflammatory arthritis with malignant lymphoma appears to be rare. Whether or not a causal relationship exists between these two entities remains uncertain.

Acknowledgment

The author wishes to thank Miss N.Malayeri for her assistance in preparing the manuscript.

Summary

A patient with malignant lymphoma who developed rheumatoid arthritis and facial edema resembling scleredema of Buschke is described. Pifalls in the clinical diagnosis of this unusual event are stressed. A possible

relationship between immunological disorders and lymphoproliferative diseases is considered.

Résumé

Le cas d'un malade atteint d'une leucémie lymphoïde qui a développé au cours de l'évolution une polyarthrite rhumatoïde est rapporté. Les relations possibles entre ces deux maladies sont discutées.

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