

LYMPHEDEMA OF THE LEG ASSOCIATED WITH RHEUMATOID ARTHRITIS

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ABSTRACT

A 14-year-old boy with a two year history of seronegative rheumatoid arthritis developed left leg lymphedema and subsequently a severe episode of lymphangitis. The diagnosis of "rheumatoid lymphedema" was confirmed by lymphoscintigraphy and conventional lymphography. Treatment consisted of bedrest and antibiotic drugs. When the signs of inflammation had subsided, therapy with corticosteroids was started with improvement of both joint pain and leg swelling. Whereas lymphedema associated with rheumatoid arthritis has been described in the upper limb of adults, to our knowledge this is the first report of the coexistent condition in the lower leg of a child.

Edema of joints affected by rheumatoid arthritis is common. In some patients, however, the edema is too generalized to be attributed solely to inflammatory synovitis. Often general factors, such as chronic anemia, hypoalbuminemia, and salt and water retention contribute to tissue swelling. Sometimes, regional complications such as venous obstruction, increased microvascular permeability or lymphatic obstruction lead to peripheral swelling of an extremity. In this report, we describe the development of lower limb edema from lymph flow obstruction in a boy with childhood rheumatoid arthritis.

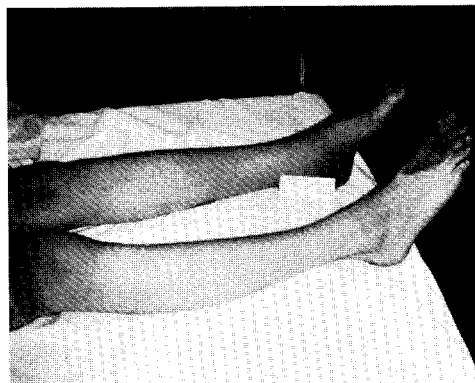


Fig. 1. Patent blue test demonstrating extensive "blue-blush" of dermal backflow into skin lymphatics of the left leg consistent with lymphedema.

CASE HISTORY

A previously healthy boy developed seronegative rheumatoid arthritis at age 12 years. The left ankle was chiefly involved with pain and swelling. Treatment was primarily with a salicylate which was initially beneficial. At 14 years of age he developed diffuse swelling of the left lower leg. Doppler examination and phlebography of the deep venous system was normal. Lymphoscintigraphy confirmed the suspicion of lymphedema.

Although initially non-operative measures for management of lymphedema were satisfactory, he presented six months later with lymphangitis, i.e., fever, swelling, pain, and dermal erythema. Erythrocyte sedimentation rate was 30mm



Fig. 2. Dorsal pedal (conventional) lymphography demonstrating defective lymphatic drainage especially in the upper left thigh and pelvis (at left upper, and above).

in the first hour. Treatment was with bedrest and IV penicillin with resolution over 3-4 days.

Patent blue injection (*Fig. 1*) showed characteristic dermal reflux and conventional lymphography (*Fig. 2*) showed discontinuity and partially obstructed lymphatics on the left side, with defective lymphatic filling in the left thigh and pelvis. Microscopic examination of the

lymph vessels on the left side revealed fibrotic obliteration.

Oral antibiotics were continued for two weeks and then salicylates were replaced by corticosteroids. The edema decreased gradually over the following months to the level present before the episode of lymphangitis.

DISCUSSION

Rheumatoid arthritis associated with peripheral lymphedema is uncommon. To our knowledge, 8 adult patients (age 43-57 years) have been reported in whom lymphedema was localized only to the upper limb (1-4). In our patient, however, the edema occurred in the lower extremity and in a 14-year-old child. As the technical studies demonstrate, the swelling is indicative of lymphedema.

The pathophysiology of the connection between rheumatoid arthritis and peripheral lymphedema, if any, is unknown. Whether rheumatoid arthritis develops in a leg with preexistent anatomical or functional lymphatic insufficiency is unclear but unlikely (3). Perhaps, as previously suggested, lymphatic dysfunction relates to byproducts released from

chronically inflamed joints (1). Apparently, however, treatment of the rheumatoid arthritis does not consistently improve the lymphedema. Accordingly, management of "rheumatoid lymphedema" remains classical non-operative therapy consisting of elastic stockinettes, limb elevation, physiotherapy, and pneumatic compression.

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