Basrah Journal of Surgery

Case Report

Bas J Surg, March, 9, 2003

LYMPHOEDEMA PRAECOX: A CASE REPORT OF PRIMARY LYMPHOEDEMA

Kathem Kassim AL-Rubiay[*] & Hussein Ali Al-Chalabi[@]

[*]DHMSA, DTMH, M.Sc, Ph.D. Consultant of Dermatology, Basrah General Hospital, [@] M.B.Ch.B., FRCP (Ed). Consultant of Medicine, Head Department of Medicine, Basrah General Hospital, Basrah; IRAQ.

Summary

A 39 year-old woman presented with clinical features which consistent with primary lymphoedema (lymphoedema praecox) associated with rosacea lymphoedema. She had clinical and radiological evidence of bilateral pleural effusion and ascites. Skin biopsy revealed perivasculitis. The treatment of this particular type of lymphoedema is extremely difficult.

Introduction

Lymphoedema praecox (LP), a type of primary lymphoedema (PL) which becomes evident at puberty occur mostly in girls and is usually confined to the lower extremities[1,2,3]. LP is caused by congenital defect in the lymphatic system, either total aplasia or partial hypoplasia [3,4,5], which lead to a low output of lymph and increased capillary filtration in that cell, proteins, lipid and debris accumalation in addition to water this results in a solid as well as a fluid component to the swelling, giving rise to the brawny nature of the oedema which does not readily pit. The predilection for limbs is due at least in part to the limited collateral drainage available at the root of limbs [2,3].

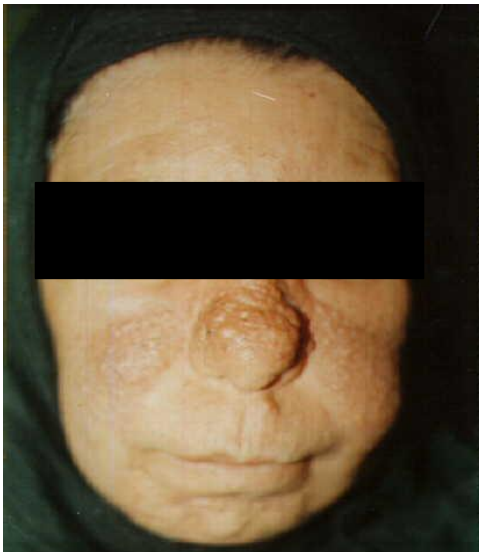
Correspondence to:
Dr.Kathem K. Al-Rubiay
Department of Dermatology, Basrah General Hospital.
P.O.Box. 289 Basrah; IRAQ.

Case Report

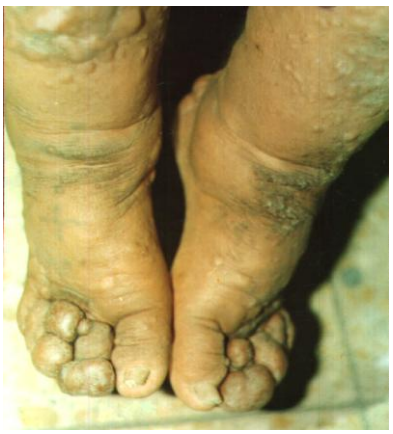
The patient was admitted to Basrah General Hospital with abdominal distention and lower extremities swelling. She had a past history of many admissions because of legs oedema. This oedema started 10 years ago as puffiness appears around the ankle and extended up ward to involve the whole legs. At first it was painful but latter become painless. She complained from discomfort, tightness and sometime itching, also erythematous skin rashes and episodic flushing mainly involving the convex area of the face.

The face had erythematous paplues, thickening and coarsening of the skin

with persistent flushing of cheeks and nose (Figure 1).



Chest examination showed sign of bilateral fluid, but the right more than left. The abdomen was distended with siagns of free fluid, with mild tenderness on deep palpation. The lower extremities were swelling with non-pitting oedema especially around ankles and toes. The skin of legs and feet was thick, hard with areas of hyperpigmented, hypertrophy and verrucous papillomata rashes around the ankles and toes (Figure 2).



Laboratory investigation were normal apart from hypoalbuinemia (total serum ablumine 1.6mg/ dL) but no albumine-urea. Radiology investigation showed bilateral pleural effusion, more on right side. Aspiration of fluid from both pleural and peritonium showed to be transudated. Ultra sound of abdomen revealed marked ascites. Skin biopsy revealed acanthotic of epidermis and blood vessel greatly increase in number and dilated with perivascular infiltration mainly by mononuclear. Lymphangio-graphy and fluorescence microlymphan-giography were not available. The diagnosis was established by careful history taking together with clinical features and histological finding. The treatment was palliative, she was put on oral antibiotic, diuretic, compression by bandage advice here to raise the legs.

Discussion

LP is the commonesr type of PL tend to affect females more frequently, it is estimated that 80% of PL will present before the age of 45 years it lead to physical, psychosocial and psycho-logical problems [2,6]. LP, in practice, simply means no identifiable outside cause to be found and it arising from an intrinsic abnormality of lymphatic system leading to accumulation of protein rich interstitial fluid and swelling of involved part especially limbs[2,3,7,8].

The major clinical changes of lymphoe-dema take place in the skin and subcutaneous tissues and such changes are of value in diagnosis. As dermal lymph stasis progress lead to dilatation of upper dermal lymphatic and accumulation of protein rich interstitial fluid, which consequent organized and fibrosis give rise to thick tight skin, latter papilliomatosis and pseudo-sclerosis[2]. Chronic facial lymphoedema is a recognized, though uncommon, complication of long-continued rosacea and may be vascular leakage and lymphatic drainage of tissue fluid in the affected skin[9].

Lymphoedema lead to episodes of secondary infection which are character-istic feature of lymphoedema. Other complications that swelling lead to

discomfort, limbs heaviness, reduced mobility, disfigurement and impaired function[2].

Lymphoedema represents end-stage failure of lymph drainage and is essentially irreversible and incurable, because of the presence of the solid component in the swelling, therefore the treatment is difficult[10]. Obvious lymphoedema is not usually a difficult diagnosis, but such cases are relatively uncommon.

References

1. Harel L, Amir J, Nussinovitch M, Straussberg Versana I. Lymphedema praecox seen as isolated unilateral arm involvement: Case report and review the literature. J Pediater 1997; 130: 492-494.
2. Mortimer PS. Disorders of lymphatic vessel. In: Champion RH, Burton JL, Burns DA, Breathanch SM, Eds. Textbook of Dermatology. 6th edition. Oxford, London, Edinburgh: Blackwell Scientific Ltd. 1998, 2277-96.
3. Daroczyl J. Pathology of lymohoedema. Clin Dermatol 1995; 13: 433-444.
4. Jansen T, Plewing G. The treatment of rosaceous lymphoedema. Clin Exp Dermatol 1997; 22: 57-59.
5. Campisi C. Lymphoedema: Modern diagnostic and therapeutic aspect. Int Angiol 1999; 8: 14-24.
6. Kirshbum M. Using massage in the relief lymphoedema. Pfr Nurse 1996; 11: 230-232.
7. Browse NL, Stewart G. Lymph oedema: Pathophysiology and classification. J Cardiovasc Surg 1985; 6: 91-106.
8. Cox NH, Paterson WD, Popple WA. Areticulate vascular abnormality in patients with lymphoedema observation in eight patients. BR J Dermatol 1996; 135: 92.
9. Scerri L, Saihan EM. Persistent facial swelling in a patient with rosacea: Rosacea lymphoedema. Arch Dermatol 1995; 131: 1071-1074.
10. Foldi E, Foldi M, Weissleder H. Conserative treatment of lymphoedema of the limbs. Angiology 1985; 36: 171-180.