

Clinical Quiz

*Abdul Q. Khan, MBBS, MS (ORTH), Mohammed-Khalid A. Sherwani, MS (ORTH), MChOrth (LIVERPOOL),
Keshav Gupta, MBBS, MS (ORTH), Somen Agarwal, MBBS, D(ORTH).*

From the Department of Orthopaedic surgery, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India. Address correspondence and reprint request to: Dr. Abdul Q. Khan, Lecturer and consultant Orthopaedic surgeon, Department of Orthopaedic Surgery, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh 202002, India. Tel. +91 9997435337. Fax. +91 (571) 2701940. E-mail: drabdulqayyum@rediffmail.com

Notice: Authors are encouraged to submit quizzes for possible publication in the Journal. These may be in any specialty, and should approximately follow the format used here (maximum of 2 figures). Please address any submissions to: Editor, Saudi Medical Journal, Armed Forces Hospital, PO Box 7897, Riyadh 11159, Kingdom of Saudi Arabia. Tel. +966 (1) 4777714 Ext. 6570. Fax. +966 (1) 4761810 or 4777194.

Lymphangiomatosis of tibia

Clinical Presentation

A 2 year old male child presented to us with complaints of swelling on whole of right lower limb since birth. Swelling was gradually progressive and the child never learnt to stand without support or walk. On and off watery discharge was present mainly near the ankle and the thigh region. The limb was swollen (**Figure 1**), soft, non tender, and local temperature was not raised and the skin yielded to the manual pressure of palpation. Blood investigations were within normal limits. X-ray of the leg (**Figure 2**) showed sclerosed and thin fibula and extensive lysis present in whole of the tibia and fibula. Fluid culture was sterile. Fine needle aspiration cytology showed abundant lymphocytes spread on lipoproteinaceous background. Abundantly dilated lymphatic spaces were present intermingled with osseous tissue.

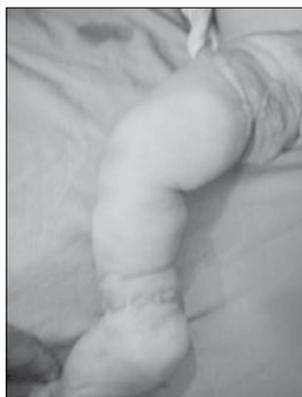


Figure 1 - Child with lymphangiomatosis affecting the right leg with lymphedema of skin.



Figure 2 - Radiograph of the leg showing extensive lysis of the tibia with thin and sclerosed fibula.

Questions

1. What is the diagnosis?
2. What are the modalities of diagnosis?
3. What is the management of this disease?

Clinical Quiz

Answers

1. Lymphangiomatosis of tibia
2. X-ray, fine needle aspiration cytology, bone scan, lymphangiography.
3. Multiple treatment modalities have been used, from radiation to corticosteroids and chlorambucil, without much success, but recently the use of interferon-alpha was found to be successful and has made it treatment of choice for this disorder.

Discussion

Lymphangiomatosis with predominant bone involvement is a rare entity. Six cases were reported so far out of which 3 had involvement of bone.¹ Most cases of lymphangiomatosis, which usually have extensive visceral involvement associated with a very poor prognosis,² however, the involvement in our variant is limited almost exclusively to soft tissues and bones of the limb and is associated with good prognosis as compared to the visceral involvement. Multiple treatment modalities have been used, from radiation to corticosteroids and chlorambucil, without much success, but recently the use of interferon-alpha³ was found to be successful and has made it treatment of choice for this disorder.

References

1. Singh Gomez C, Calonje E, Ferrar DW, Browne NL, Fletcher CDM. Lymphangiomatosis of the limbs: Clinicopathologic analysis of a series with a good prognosis. *Am J Surg Pathol* 1995; 19: 125-133.
2. Hilliard RI, McKendry JB, Phillips MJ. Congenital abnormalities of the lymphatic system: a new clinical classification. *Pediatrics* 1990; 86: 988-994.
3. Reinhardt MA, Nelson SC, Sencer SF, Bostrom BC, Kurachek SC, Nesbit ME. Treatment of childhood lymphangiomas with interferon-alpha. *J Pediatr Hematol Oncol* 1997; 19: 232-236.

www.smj.org.sa

Saudi Medical Journal Online features

- * Instructions to Authors
- * Uniform Requirements
- * STARD
- * Free access to the Journal's Current issue
- * Future Contents
- * Advertising and Subscription Information

All Subscribers have access to full text articles in HTML and PDF format. Abstracts and Editorials are available to all Online Guests free of charge.