Clinical Quiz

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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.

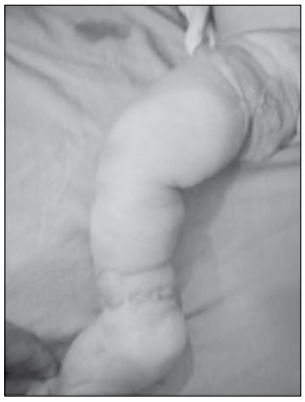

Figure 1 - Child with lymphangiomatosis affecting the right leg with lymphedema of skin.

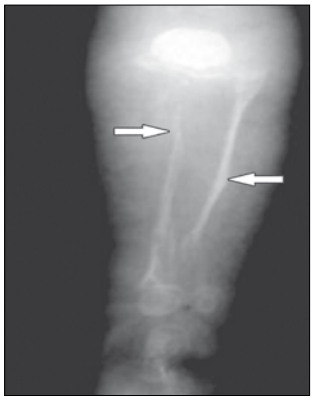

Figure 2 - Radiograph of the leg showing extensive lysis of the tibia with thin and sclerosed fibula.

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-alpha[3] was found to be successful and has made it treatment of choice for this disorder.

References

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