ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

https://iap-kpj.org

Karnataka Paediatric Journal

Article in Press

Case Report

Presentation of lymphoedema praecox as a septic shock with acute renal failure in an adolescent girl: A case report

Rajendra Raghunath Khadke[1], Abhijit Venkatesh Joshi[2], Prachi Samidh Patel[1]

[1]Department of Pediatrics, Varad Medical Foundation, [2]Department of Pediatrics, Chiranjeev Children's Hospital and Critical Care Center, Aurangabad, Maharashtra, India.

***Corresponding author:**
Abhijit Venkatesh Joshi, Department of Pediatrics, Chiranjeev Children's Hospital and Critical Care Center, Aurangabad, Maharashtra, India.

joshiabhijit71@yahoo.com

Received: 21 June 2023
Accepted: 30 July 2023
EPub Ahead of Print: 15 September 2023
Published:

DOI
10.25259/KPJ_42_2023

Quick Response Code:

ABSTRACT

Lymphoedema praecox (Meige disease) is a rare chronic disease of disordered lymphatic transport in which 10% of the cases present as non-inherited mutations responsible for defective lymphatic development. The inherent complex inflammatory pathways lead to defective lymphangiogenesis, oedema, adipose tissue deposition and chronic fibrosis. Various factors lead to local impairment of adaptive immunity leading to the increased incidence of bacterial infections. Sepsis and septic shock arising from such infection can be life-threatening. One such case is discussed where a post-pubertal adolescent girl presenting with a chronic painless unilateral limb swelling landed into cellulitis and gangrene with septic shock. She needed care in an intensive care setting and a diagnostic work-up was started to look into the cause. She recovered from this complication and care for this chronic condition was continued later. A high index of suspicion for the occurrence of this rare entity with its associated complications is the prerequisite to a successful outcome.

Keywords: Lymphoedema, Late onset, Hereditary, Meige lymphoedema

INTRODUCTION

Lymphoedema, a chronic disease of a disordered lymphatic transport, consists of primary lymphedema, which is rare (incidence 1.15 in 100,000 in age <20y population) and is differentiated into hereditary Type 1 (Milroy disease), Type 2 (lymphoedema praecox or Meige disease) and lymphoedema tarda. Primary lymphoedema almost always affects the paediatric population; adult-onset disease is uncommon. Boys are more likely to present in infancy, while girls commonly develop the disease in adolescence. The primary disease affects the lower extremities in 93% of cases. Lymphoedema praecox is seen commonly in post-pubertal adolescent girls with involvement of the left lower limb as the common site.

Patients may present with swelling over the limbs, paraesthesias and poor healing of wounds. The assumed hypothesis includes the local proliferation of T regulatory cells causing impairment of adaptive immunity.[1] This, along with the loss of dendritic cell function and a breach in the epidermal-dermal layer, provides entry for the infection.[2] It may present with systemic signs of high fever and rigors and local signs of erythema, pain, warmth and swelling. Beta-haemolytic streptococci can cause local cellulitis, lymphangitis and sometimes gangrene. Septic shock may ensue, if not suspected and treated in time and can be life-threatening. This case report

stresses the need for a timely diagnosis of this rare disorder of the lymphatic system and anticipation of the associated complication of bacterial sepsis which may give rise to a catastrophe like acute renal failure. It may also sensitise the treating physician about the need for long-term care of this chronic condition and about the education of the patient and the family for care at home. They are also taught about seeking help at any possibility of complications.

CASE REPORT

We have discussed a case of a 16-year-old post-pubertal girl from a rural background. Her presenting complaints were fever, swelling, pain and ulceration involving the dorsum of the left foot over 4 days. She also started vomiting and passing loose stools. She presented to the emergency department in septic shock and acute renal failure. Her parents gave an account of her having similar complaints around 2 years back with swelling over her left foot and ankle and needing hospitalisation for 8 days. She recovered from the illness with the treatment but painless left leg swelling persisted for 2 years and discomfort during walking continued. She was worried about the discomfort while walking and doing daily chores, particularly changing clothes. No specific help or any treatment was sought for this purpose and failed to follow up with the hospital where she was previously treated. Her health seemed normal except for the persistent painless leg swelling. Her menstrual history revealed that she attained menarche at 12 years but had irregular cycles and sometimes heavy and prolonged menstrual bleeding. She was born of a non-consanguineous marriage. Her family history did not reveal any significant illness.

Her physical examination showed a non-haemorrhagic blister over the dorsum of the left foot and gangrenous skin changes over the great toe and the adjacent toes [Figure 1]. There was non-pitting limb oedema and erythema to the middle third of the leg. Stemmer's sign was positive which was elicited by pinching the skin over the dorsum of the base of the second toe.[3]

Figure 1: Non-haemorrhagic blister over the dorsum of the left foot and limb oedema with skin gangrene over the great toe and adjacent toes.

Her emergency care in Intensive Care Unit started with the management of septic shock with fluid support, inotropes and antibiotics. She responded to the treatment with recovery from shock and acute renal failure. Ulceration and gangrene were treated simultaneously with the local care of the wound with regular dressing and wound debridement.

Her deranged laboratory parameters were anaemia and raised inflammatory markers and raised serum creatinine. She underwent Doppler of the limb which showed pan reversal of diastolic flow in the left lower limb arteries possibly secondary to systemic infection. Ultrasound of the abdomen showed medical renal disease and echocardiogram was normal. Computed tomography angiography of the limb did not show changes of vasculitis. Her antinuclear antibody was negative. Lymphoscintigraphy could not be done due to a lack of expertise and experience in doing the test.

During recovery, measures were taken like giving compression dressing to the affected limb and leg elevation to decrease limb oedema.[4] She was counselled about the care of the limb with the use of elastic garments[5] and family members asked to keep a watch on new infections, if any.

DISCUSSION

The diagnosis, in this case, is primary lymphoedema looking at the age at presentation in a post-pubertal adolescent girl, the chronicity of painless limb swelling and no risk factors for chronic venous insufficiency. The unilateral firm, sclerotic skin with non-pitting oedema and a positive Stemmer's sign ruled out other diagnoses clinically. Stemmer's sign is more sensitive than specific. If the test is positive, it is likely that the patient has lymphoedema. The staging was done as Stage 3 in view of fibroadipose tissue deposition and the associated skin changes. Causes of secondary type such as filariasis, trauma and malignancy were ruled out by relevant investigations.[6] Secondary lymphoedema is uncommon in children but is responsible for the disease in 99% of adults. This case report highlights the possibility of this rare non-inherited disorder in an adolescent female wherein the primary symptoms like swelling may not prompt the patient to seek medical help. Furthermore, this condition may be missed by the attending physician due to the probability of being unaware about this condition. Children with suspected primary lymphoedema need to be examined for syndromic characteristics that are associated with the disease. The catastrophe of sepsis and associated poor outcome can be avoided if dealt properly with this entity. For a favourable outcome in such a rare condition with the associated complication of sepsis, a high

index of suspicion, gained from knowledge and awareness on the part of the treating physician is highly required. This case report may sensitise the paediatrician to deal with such a catastrophe of this intriguing disorder.

CONCLUSION

Lessons learnt are:

1. Lymphoedema is to be differentiated from other types of oedema clinically and staging is also possible with the help of a positive Stemmer's sign
2. Primary lymphoedema is a clinical diagnosis made only after ruling out other possible causes of lymphoedema
3. A high index for sepsis as a possible complication should be kept to prevent a catastrophe.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Duhon BH, Phan TT, Taylor SL, Crescenzi RL, Rutkowski JM. Current mechanistic understandings of lymphedema and lipedema: Tales of fluid, fat, and fibrosis. Int J Mol Sci 2022;23:6621.
2. Nores GD, Ly CL, Savetsky IL, Kataru RP, Ghanta S, Hespe GE, *et al*. Regulatory T cells mediate local immunosuppression in lymphedema. J Invest Dermatol 2018;138:325-35.
3. Kamijo E, Ishizuka K, Shikino K, Sato E, Ikusaka M. Physical findings and tests useful for differentiating lymphedema. J Gen Fam Med 2021;22:227-8.
4. Vignes S, Albuisson J, Champion L, Constans J, Tauveron V, Malloizel J, *et al*. Primary lymphedema French National Diagnosis and Care Protocol (PNDS; Protocole National de Diagnostic et de Soins). Orphanet J Rare Dis 2021;16:18.
5. Senger JB, Kadle RL, Skoracki RJ. Current concepts in the management of primary lymphedema. Medicina (Kaunas) 2023;59:894.
6. Borman P. Lymphedema diagnosis, treatment, and follow-up from the view point of physical medicine and rehabilitation specialists. Turk J Phys Med Rehabil 2018;64:179-97.

How to cite this article: Khadke RR, Joshi AV, Patel PS. Presentation of lymphoedema praecox as a septic shock with acute renal failure in an adolescent girl: A case report. Karnataka Paediatr J, doi: 10.25259/KPJ_42_2023