Case Report

Primary lymphedema of the left lower limb in a new-born boy: a case report

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ABSTRACT

A 7-day-old boy was admitted with a history of swelling involving his left lower limb since birth. He was born normally to a Gravida 1, Para-0, to a 22-years-old mother. Antenatal history was nothing significant. His clinical examination revealed left lower limb swelling and was suggestive of primary lymphedema. It involved from the ankle area up to the groin of the left side of the limb. His right lower limb and both upper limbs were normal. His other systemic examination was also normal. He was treated with aseptic needle aspiration of the lymphatic fluid, and a compression dressing was applied. He suddenly died of an unknown reason while sleeping at around 4.00 AM on the third post aspiration day.

Keywords: Case report, Congenital anomaly, Cystic swelling, Lymphedema, Primary lymphedema

INTRODUCTION

Primary lymphedema results from congenital abnormality or dysfunction of the lymphatic vessels. Secondary lymphedema is more common than the primary form can develop as a consequence of destruction or obstruction of the lymphatic channels by other pathological conditions such as infection, trauma, or malignancy.

Lymphoscintigraphy, Single-photon emission computed tomography (SPECT), computed tomography (CT), and Magnetic resonance imaging (MRI) lymphangiography are the radiological investigation offered for the diagnosis and confirmation of lymphedema.

The options for the management of lymphedema are compression therapy, physiotherapy, manual lymphatic drainage, and operative therapy. Surgical CAse REport (SCARE) 2020 guidelines published by Agha et al are followed for reporting the present case.

CASE REPORT

A 7-day-old boy was admitted to the department of pediatric surgery with a history of swelling involving his left lower limb since birth. He was full-term, delivered at hospital normally, to a Gravida 1, Para-0, 22-years-old mother. Antenatal history was nothing significant and antenatal check-up was irregular. Antenatal sonography done at the 7th month reported that baby had swelling over the left leg/limb. Family history also did not detect any significant maternal or paternal families. He was 2.5 kgs, his general and systemic examination was also normal. He was treated with aseptic needle aspiration of the lymphatic fluid, and a compression dressing was applied. He suddenly died of an unknown reason while sleeping at around 4.00 AM on the third post aspiration day.

His left lower limb examination revealed a huge soft, cystic, non-tender swelling involving the entire parts of the limb. The swelling extended from just above the ankle area to the inguinal area (Figure 1A, 1B). The skin over the swelling was discolored and irregular. A transillumination test was done which revealed that the swelling was positive at all the areas of the limb and the
thigh (Figure 2 and B). His right lower limb and both the upper limbs were normal. Other systemic examinations were also normal. Skiagram of the left lower limb reported as normal for bony structures (Figure 3A). The affected left lower limb’s ultrasound was reported as a multi-septated, cystic lesion (Figure 3B). Doppler study of the left lower limb was tried, but could not be done due to the multi-septated lesions, and the details of the vessels of the affected limb could not be revealed. Venous anatomy of the affected limb appeared normal, but spectral patterns could not be ascertained.

His haematological investigations were as; Hb 18.6 g/dl, White blood cell count 5000 /mm3, total bilirubin was 8.2 mg% (direct 1.25 mg% and indirect 6.95 mg%), SGOT/SGPT, and ALP was within normal limits. His serum proteins reported as 4.5 gm% (albumin 2.73 gm%, globulin 1.77 gm% and A/G ratio was 1.54.

The baby was consulted with the plastic surgeons and advised needle aspiration of the swelling due to the cystic nature. All details of the procedure, requirements of major surgical interventions in future, and the morbidity and mortality relating to the disease were explained to the relatives. After obtaining informed consent, under all the aseptic precautions, needle aspiration of the swelling was done and approximately 250 ml of golden yellow fluid was aspirated (Figure 4A), and the same was reported as lymphatic fluid on biochemical analysis. After needle aspiration of lymphatic fluid, marked reduction in the size of the swelling of the affected lower limb was observed (Figure 4B-D).

His packed cells transfusion was given for the treatment of hypoalbuminemia. He was doing well and accepting feeds well in the postoperative period, but suddenly at early in the morning at around 4.00 AM, the mother reported that the baby was cyanosed and not taking
respiration. He died due to respiratory failure and could not be revived.

DISCUSSION

Lymphedema is an accumulation of protein-rich fluid in the interstitial space due to the reduction in the capacity of lymph transport.1,2 Lymphedema is classified as primary and secondary lymphedema.1,2 Primary lymphedema is congenital in origin and it results from the abnormal or faulty development of the lymphatic channels. There may be aplasia, hypoplasia, or dysplasia of the lymphatic channels/system, and results in the accumulation of the lymphatic fluid at the affected parts.1-5 Secondary lymphedema is acquired in nature, and it is due to a consequence of destruction or obstruction of the lymphatic channels by other pathological conditions such as infection, trauma, post-operative, malignancy, or post-radiation. Secondary lymphedema is more common than primary lymphedema.1,2,4

The prevalence of primary lymphedema is approximately 1.15/100,000.13 In children, primary lymphedema is more common than secondary lymphedema. Depending upon the age of clinical presentation, primary lymphedema is categorized congenital, praecox, and tarda. It is called “congenital” when it occurs before the first 2-years of life. It is called “praecox” when it occurs 2-years onwards but before the 35-years. It is called “tarda” when it occurs after the age of 35-years.2,13 In approximately 90% of the cases, primary lymphedema involves the lower limbs. Other sites are upper limbs, it may be unilateral or bilateral involvements. In some cases, primary lymphedema may be a part of other malformations.3,4,9

Lymphoscintigraphy is frequently performed, is the safe and reliable investigative procedure for the diagnosis and confirmation of lymphedema.6 Single-photon emission computed tomography / computed tomography (SPECT/CT) is also helpful in the evaluation of lymphedema. The addition of SPECT/CT scan also provides additional information in the form of the extent of the disease, status of lymph nodes, etc.7 Magnetic resonance imaging lymphangiography is also helpful in the evaluation of cases of primary lymphedema.8

There are no known therapeutic agents that cure or treat lymphedema. The objectives of the management for lymphedema are reduction in the volume of lymphatic fluid, preventing the occurrence of complications associated with lymphedema, and improving the quality of life of patients. In general, the therapeutic options for managing lymphedema cases are compression therapy, physiotherapy, exercise, manual lymphatic drainage, skin care, and operative therapy. The surgical therapy offered for the management of lymphedema is volume reduction / cutaneous resection, liposuction, and reconstructive operative procedure as lymphovenous anastomosis. If surgical therapy is indicated and required, lymphovenous anastomosis is the most common surgical procedure done for the same. Active involvement of parents is most important in the care of children diagnosed with primary/congenital lymphedema.3,9,11,13,14

The present case was clinically diagnosed as a case of primary congenital lymphedema of the left lower limb and was subjected to needle aspiration of lymphatic fluid and compressing dressing. Unfortunately, he died prior to completion of the therapy offered to him, and therefore the result of the same could not be evaluated.

CONCLUSION

Primary congenital lymphedema may involve the entire limb and disfigure the same. For the cases presenting with a large volume, and cystic lymphedema during infancy, a needle aspiration with compression dressing may be offered as an initial therapeutic option for the management.

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