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Case Report

Paediatrics Section

Effective Role of Oral Sildenafil in the Treatment of Cystic Hygroma in an Infant: A Case Report

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ABSTRACT

Cystic hygroma is a benign congenital malformation of the lymphatic system. Most cystic hygromas are found in the neck; rarer locations include the axilla, mediastinum, and limbs. Symptoms range from incidental findings to significant morbidity due to compression of adjacent organs, infection, haemorrhage, etc. Treatment is primarily aimed at complete surgical resection. Other treatment modalities include sclerotherapy, radiotherapy, laser ablation, and medical therapy with sirolimus, but recurrence rates are high. Surgery is not always possible since Lymphatic Malformations (LMs) can be intertwined within muscles or organs, and incomplete resection of LMs can result in recurrence; hence, alternative therapies have been explored. Recently, authors reported a case of a full-term female infant with a left-sided cystic hygroma. There was marked regression of the cystic hygroma with oral sildenafil, a selective inhibitor of phosphodiesterase-5.

Keywords: Lymphangiomas, Lymphatic malformations, Sclerotherapy

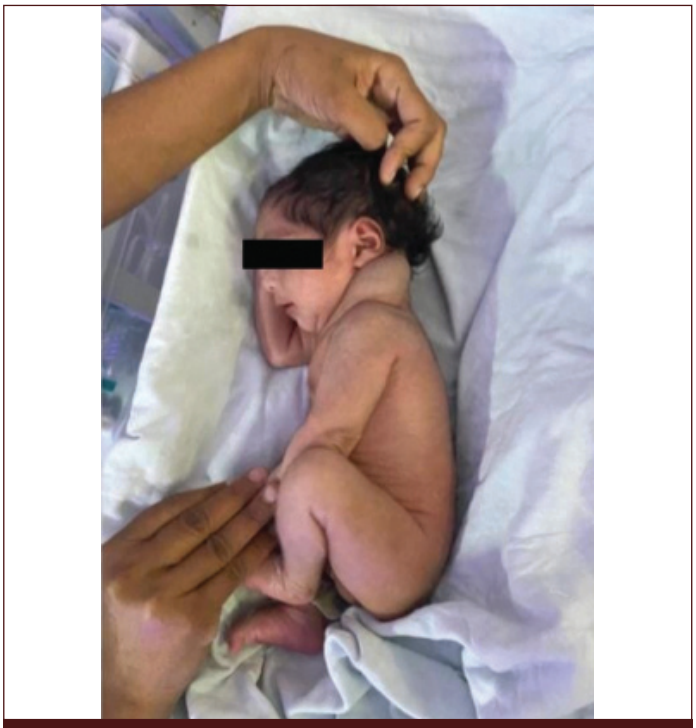
CASE REPORT

A full-term female infant weighing 2500 g was delivered vaginally to a 28-year-old mother at Department of Neonatology, Government Medical College (GMC), Aurangabad, Maharashtra, India. Delayed cord clamping after one minute was performed. Appearance, Pulse, Grimace, Activity, Respiration (APGAR) scores were 8 and 9 at one and five minutes, respectively. A swelling (approximately 10×4 cm) was noted on the left-side of the neck and back. The mass was antenatally detected on prenatal ultrasound in the third trimester at 33 weeks, which showed cervical meningocoele (52×39 mm cystic lesion with septations). At birth, the baby weighed 2500 g and had a head circumference of 33 cm. The vitals are shown in [Table/Fig-1]. The baby was initially fed Expressed Breast Milk (EBM) via a spoon every three hours, followed by breastfeeding. No signs of respiratory distress were noted on admission.

Parameters	Findiings
HR	132/min
CFT	<3 sec
Colour	Pink
RR	40/min
BP	62/46 (mmHg) (MAP ≈51 mmHg)
SpO_2	>95% at room air (preductal and postductal)
Anterior fontanelle	Normal pulsatile
P/A	Soft, no organomegaly

[Table/Fig-1]: The vitals of the neonate.
HR: Heart rate; CFT: Capillary refill time; RR: Respiratory rate; BP: Blood pressure; MAP: Mean arterial pressure; P/A: Palpation of the abdomen

Local examination of the neck swelling: On inspection, a swelling was present on the left-side of the posterior triangle of the neck, with a horizontal dimension of 10 cm and a vertical dimension of 4 cm, having a rounded shape, sharp margins, and a smooth surface. The overlying skin showed slight bluish discoloration with normal surrounding tissue. On palpation, there were no signs of inflammation or tenderness. The mass was soft, cystic, and compressible with fluctuation. Translucency was absent; it was not reducible and non-pulsatile. Mobility was not restricted [Table/Fig-2].



[Table/Fig-2]: Cystic hygroma before treatment at birth.

Diagnosis: Ultrasound of the lesion showed a multicystic lesion with internal septations and no blood flow detected on colour Doppler ultrasound, suggestive of cystic hygroma. Paediatric surgeon consultation was obtained for further evaluation and treatment. Magnetic Resonance Imaging (MRI) of the neck could not be performed due to the patient's family's financial constraints.

Treatment: Since the patient's parents did not consent to surgical management, the alternative option of medical management with sildenafil was started after informed consent was obtained.

At baseline (day 6), treatment was initiated with sildenafil (Vigorex 25 mg) at 1 mg/kg per dose, administered three times daily orally mixed with milk [1]. Baseline physical examinations were performed, and vital signs, including blood pressure, heart rate, respiratory rate, oxygen saturation, and basal body temperature, were obtained before initiating sildenafil. After dispensing the initial dose, vital signs were monitored every 30 minutes during a 2-hour observation period. The neonate remained well and on breastfeeding during her stay in the neonatal unit.

The neonate was given sildenafil thrice daily for 20 weeks. The swelling did not show any regression one month after treatment [Table/Fig-3]. The infant was evaluated in the clinic at weeks 4, 8, 12, and 20 and was contacted by phone in between. At week 20 (end of treatment), sildenafil was discontinued. The infant was observed for 12 weeks after the last dose and followed up till one year of age. The swelling began regressing at eight weeks (2 months) of treatment [Table/Fig-4-7]. Outcomes included both the physician's and parents' assessments of swelling improvement compared with baseline. At each visit, the size of the swelling, changes in texture, and overall change compared with baseline were recorded [Table/Fig-8]. No changes in texture were noted. Photographs were taken for comparison. Parents were asked to review and report any adverse events. No specific side effects were observed during the treatment course. Parents remain in regular outpatient follow-up; no untoward effects have been noted so far.

[Table/Fig-3]: After one month treatment (No regression of swelling).

[Table/Fig-4]: After two months of treatment (Partial regression of swelling). **[Table/Fig-5]:** After four months of treatment (90% regression of swelling). (Images from left to right)

[Table/Fig-6]: After five months of treatment (Complete regression of swelling). **[Table/Fig-7]:** Baby at one year of age (no recurrence). (Images from left to right)

Age of the baby	Size of lump
At birth	Approximately 10x4 cm
4 weeks	10×4 cm
8 weeks	8×3 cm
12 weeks	6×3 cm (swelling was soft and its borders were difficult to identify)
16 weeks	2×1 cm (90% regression)
20 weeks	No swelling, only loose skin fold
1 year	No recurrence

[Table/Fig-8]: Change in the size of the lump with the age of the neonate.

DISCUSSION

The patient in the present report was treated with sildenafil (Vigorex 25 mg) orally as 1 mg/kg per dose thrice daily for 20 weeks, in the same dosages as those in the study by Ullah MS et al., [1]. Cystic LMs are localised areas of abnormal development of the lymphatic system [2]. Several mechanisms have been proposed to explain the pathophysiology of cystic hygroma. They can be categorized based on cyst size as microcystic, macrocystic, and mixed lymphangiomas [3]. Cystic hygroma, a macrocystic lymphangioma, occurs more frequently than other types of lymphangiomas and may be composed of single or multiple macrocystic lesions with limited communication with normal lymphatic channels [4]. The term is usually used for congenital LMs detected in utero or observed at birth. Diagnosis is commonly made clinically based on large size, location, and translucence. Although it tends to enlarge progressively over months, a relatively rapid increase in size has also been described [5]. Patients can present with visible deformity, pain related to compression of adjacent structures, or sudden enlargement of LM due to haemorrhage or infection [5,6].

The management of cystic hygroma is preferably surgical. Indications for surgery in paediatric cases include significant cosmetic deformity, obstructive symptoms, bleeding, and recurrent infections [4,7]. Other treatment modalities include aspiration, radiation, and injection of sclerosants such as bleomycin and OK-432, derived from a strain of Streptococcus pyogenes [8]. Lymphangiomas have also been successfully treated with rapamycin (sirolimus) [9,10].

Sclerosants such as ethanol, doxycycline, bleomycin, and OK-432 are less effective for treating microcystic and mixed lesions. Although macrocystic lesions may respond well initially, patients often require repeated sclerotherapy treatments throughout their lifetime [5,11]. The efficacy of oral medications for the treatment of LMs, including sirolimus and propranolol, requires further investigation [10,12].

Sildenafil selectively inhibits phosphodiesterase-5, preventing the breakdown of cyclic Guanosine Monophosphate (cGMP). Inhibition of phosphodiesterase-5 decreases the contractility of vascular smooth muscle, producing vasodilation [13]. A potential explanation for the therapeutic effect seen in these cases is that relaxation of the lymphatic vasculature may allow lymphatic spaces to open, thereby decreasing LM volume. Similar cases and studies reported in the literature has been depicted in [Table/Fig-9] [1,14,15].

Authors name	Median pretreatment size of the lesion	Median post-treatment size of the lesion	Adverse effects
Ullah MS et al., [1]	13.30 cm^2	0	21.82% (present)
Swetman GT et al., [15]	Lymphatic anomalies significantly improved (75% reduction in size in one subject)		No
Danial C et al., [16]	6 of 7 patients showed therapeutic response, 4 subjects had lymphatic volume decrease (1-31.7%), clinical improvement was noted in 2 subjects despite an increase in lymphatic volume (1.1-3.7%)		Mild
Present study	10×4 cm	0 cm	No

[Table/Fig-9]: Types of implant used for internal distraction plating in distal radius fractures [1, 15 16].

CONCLUSION(S)

In present case, oral sildenafil produced excellent size reduction in cystic hygroma and no side effects. Additionally, it was easy to administer and quite affordable compared with surgical intervention. Considering the effectiveness of sildenafil in this case, the drug may be added as another effective agent in the range of treatment modalities for cystic hygroma, despite the highly limited data in the literature about this off-label indication. Further studies with larger sample sizes and long-term follow-up are advised to assess the efficacy and safety profile of oral sildenafil.

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