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Chin Aun Low, Foead Agus Iwan

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Case Report: We report a newborn baby with Down's syndrome presented with cystic hygroma in his left arm and forearm. Patient was treated with OK-432 for the swelling of the left arm. However, patient presented at five months of age with severe sepsis secondary to gangrene of left arm and concomitant meningitis which subsequently led to his death.

Conclusion: Picibanil aka OK-432 has been increasingly used for the last two decades for the treatment of cystic hygroma with much success. However, few literatures report of serious morbidity with its use. More case controlled studies should be carried out to further understand the side effects of sclerosant therapy, as it has the potential to be ideal treatment for cystic hygroma in the future.

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CASE REPORT

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Keywords: Cystic hygroma, Lymphangioma, Picibanil, Neonatal gangrene

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INTRODUCTION

Cystic hygroma is the outcome of congenital developmental failure of lymphatic system in the body.

Most cases of cystic hygroma reside in the head and neck region. Other sites are superior mediastinum, axilla, retroperitoneum, mesentery, pelvis and lower limbs. Its occurrence in upper extremity is scarcely reported in literature. Approximately, 50% of cases are diagnosed at birth [1]. Since the introduction of sclerosant therapy, a non-surgical method of treatment has been favored over surgical treatment. We report a newborn with Down's syndrome presented with cystic hygroma in the arm and forearm. He was subsequently treated with a sclerosant known as Picibanil (OK-432) which is a mixture originating from group A *Streptococcus pyogenes*.

CASE REPORT

A newborn male neonate with features of Down's syndrome presented to the special care nursery (SCN) to our institution with complaint of swelling of left arm and left forearm on day-1 of life. There were no other complaints. Clinical examination revealed swelling of the left neck extending to the left arm until proximal forearm measuring 12x8x7 cm. The swelling showed no sign of inflammation. On examination, the swelling appeared to have multiple cystic areas, which were easily compressible and brilliantly transilluminant without any bruit.

Magnetic resonance imaging (MRI) scan revealed cystic lesion extending from C2 proximally to left

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proximal forearm distally. The upper neck lesion appeared subcutaneous and lower neck lesion extended into the intermuscular plane of left sternocleidomastoid, infraspinatus and deltoid. At the chest level, it lied between left pectoralis major and pectoralis minor anteriorly and extended up to left teres major and latissimus dorsi posteriorly. At the arm level, it compressed the left coracobrachialis muscle (Figure 1). Neurovascular structures were intact and normal bone structures were shown.

Our diagnosis was cystic hygroma of left arm and left forearm that was evidently supported by MRI. The patient was then referred to a tertiary center and was treated with OK-432 sclerotherapy. Fine-needle aspiration was not done to prevent introduction of infection to the upper limb and due to high recurrence rates of lymphangioma after aspiration as a therapeutic procedure.

The baby started the OK-432 therapy at third months of life. The patient was given a single dose of 0.2 mg (20 mL) of OK-432 over the left deltoid where the swelling was most prominent. There was noticeable reduction in the size of the swelling for the first two months from time of injection to the size of 8x5.5x4 cm. However, he was admitted to SCN in our institution at fifth month with complaints of fever, fits and blackish discoloration of the left upper limb. The parents denied any traditional medication applied to the baby's left arm and forearm. On examination, patient has pyrexia and there were gangrenous patches over the left arm which was the site of previous injections of OK-432 with an open wound measuring 6x2 cm over the lateral aspect of the left arm (Figure 2). The patient was then intubated in view of severe metabolic acidosis with pH of 6.73 and was supplemented with intravenous bicarbonate. The patient was treated as severe sepsis secondary to gangrene of left upper limb and concomitant meningitis. The patient was started on dual antibiotic therapy with intravenous ampicillin and cefotaxime given based on weight-based dosage. On day two of admission, patient was diagnosed with disseminated intravascular coagulation and was transfused with fresh frozen plasma, platelet and packed cells. 500 cm³ of hemoserous fluid was aspirated from the left upper limb as a therapeutic procedure but the swelling recurred within the following two days. Unfortunately, the baby's condition deteriorated over the following five days and died due to unresolved severe sepsis.

DISCUSSION

Lymphangiomas may be classified histologically into three types namely simple, cavernous, and cystic. Simple lymphangiomas are composed of minute lymphatic channels communicating with its stroma components whereas cavernous lymphangiomas consists of dilated lymphatic components surrounded by stroma. Cystic lymphangioma are larger cystic lesions which poorly communicate with the lymphatic elements.

Figure 1: Magnetic resonance imaging (MRI) scan showing cystic lesion extending from C2 proximally to left proximal forearm distally.

Figure 2: Severe infection of left upper limb with blistering and necrotic patches seen.

Although cystic hygroma has been associated with a nuchal lymphangioma, Turner's syndrome, and Noonan syndrome, there is evidence that cystic hygroma occurs more frequently in Down's syndrome. It has been postulated significant correlation with Down's syndrome as patients with Down's syndrome have potentially enlarged jugular lymphatic dilatations [2].

Traditionally, cystic hygroma has been treated with surgical excision. However, this mode of treatment was often accompanied with serious complications, namely infection, recurrence, bleeding and injury to major nerve

and vessels. Surgical excision was only accompanied with a one-third possibility of successfully preserving all vital structures [3]. The recurrence rates are high even after apparent complete excision of the lesion.

Sclerotherapy as the primary modality of treatment is under trial in many centers. The sclerosant agents currently under researches are: OK-432 (Picibanil), bleomycin, doxycycline, alcohol, alpha interferon-2 and fibrin sealant [4]. Most of the studies documented complete resolution of the lesions with OK-432 in about 60–80% of cases.

OK-432 (Picibanil) is a lyophilized incubation mixture of group A *Streptococcus pyogenes* of human origin. Since its introduction as the main therapy for lymphatic malformations by Ogita in 1987, it claims better results with fewer complications. OK-432 acts by provoking inflammatory responses towards inactive bacteria, leading to increased endothelial permeability, which increases lymphatic drainage of that selected cystic region. In the later stage, fibrosis of cystic spaces predominates and will lead to shrinkage of the tumor [5].

Poldervaart's study showed that microcystic variety of lymphatic malformation treated with OK-432 alone had 27% of excellent result, 33% good result and 40% poor result, while of macrocystic variety of lymphatic malformation; 88% had excellent result. Since cystic hygroma is of macrocystic variant, the expectation of successful therapy with OK-432 alone is above 80%. In their data, the recurrence rates vary from 5–8%. The adverse effects are mostly mild, such as mild edema, erythema, pyrexia, induration and wrinkling of skin at site of injection [6]. Almost all of the adverse effects disappear after a week.

We presume the open wound and gangrene was due to unresolved infection at the site of injection, but we cannot be certain whether it was attained at time of injection or subsequent poor wound care or secondary infection caused by underlying tissue inflammation. It is obvious that the death of the patient was due to severe sepsis. Whether there is any correlation with the injection of OK-432 cannot be clarified. Most of the trials of OK-432 have short follow-up. There are uncertainties when it comes to cure and regression. Furthermore, mortality rate associated with the use of OK-432 has never been reported.

CONCLUSION

Cystic hygroma of left arm and left forearm is rarely reported. Picibanil aka OK-432 has been increasingly used for the past 2 decades for the treatment of cystic hygroma with much success. However, few literatures report of serious morbidity with its use. More case controlled studies should be carried out to further understand the side effects of sclerosant therapy, as it has the potential to be the ideal treatment for cystic hygroma in the future.

Author Contributions

Chin Aun Low – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Foead Agus Iwan – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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