Management of cystic lymphangiomas in Ile-Ife, Nigeria

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Abstract

Background: The management of cystic lymphangiomas is and challenging. Of all the available modalities of treatment, surgery remains the gold standard but it is associated with significant morbidity and mortality.
Method: Retrospective analysis of 28 cases of cystic lymphangioma seen at the Obafemi Awolowo University Teaching Hospital, Ile-ife, Nigeria.
Results: There were 16 boys and 12 girls with a median age of 6 months. The cervical region was most commonly involved site in 19(67.9%) children. Eight (28.6%) of the patients had morbidity at presentation. Only 24 of the 28(86%)patients had surgical excision. Complete macroscopic excision was possible in 15 (62.5%) patients. There was one death (4.2%) at induction of anaesthesia due to rupture of a cervical cyst at intubation with aspiration. Post-operative complications occurred in 10 (41.7%) patients. Recurrence was reported in 1(4.6%) patient.
Conclusion: The presentation of cystic lymphangiomas in our own environment is diverse. There is a high preoperative morbidity. Surgery remains the only option available in this environment but is associated with a high e complication rate.

Key words: Cystic lymphangioma, morbidity, mortality

Introduction

Cystic lymphangiomata (CL) are congenital malformations of the lymphatic system appearing as single or multi-loculated fluid filled cavity mainly in the cervical or axillary region but could also be found in other rare sites. [1, 2] Embryologically, they are thought to be due to sequestration of a portion of the primitive lymphatic anlage. Surgery is regarded as the first choice treatment but because of the dangers associated with conventional surgical excision especially in the cervical CL, other modalities of treatment such as the use of bleomycin emulsion, intra-lesional OK 432, intra-lesional triamcinolone and fibrin sealant

are being tried. [3-6] These treatment modalities are not available in our own environment hence surgery remains the only line of treatment available. Surgery may be complex and multiple and may need to be staged to allow complete excision of the lesion. Incomplete excision may be followed by recurrence of the lesion.

We report our experience with the management of cystic lymphangiomas at the Obafemi Awolowo University Teaching Hospital in Ile-Ife, southwestern Nigeria.

Materials and methods

This is a retrospective study of all children with cystic lymphangiomas seen between 1978 and 1998 at the Obafemi Awolowo University Teaching Hospital, a semi urban teaching hospital in Ile Ife Nigeria. The clinical records of the patients were reviewed for age, sex, clinical presentation, location and distribution of the lesions as well as the outcome of treatment. All the patients had radiological examination while ultrasound was added later when it became available. Additional investigations such as intravenous urography, barium meal/enema were used for further evaluation where intra-abdominal cystic lymphangioma were suspected to be present

Results

Twenty-eight children with cystic lymphangioma were seen during the study period. There were sixteen males and twelve females with age ranging from 1 day to 13 years (median age of 6 months). The age and the distribution of the lesions is shown in table 1.

Eighteen (64.3%) of the children were less than 1 year at presentation. Seventy five percent of the children presented before two years of age. In 16 (57.1%) children, the lesions were noted at birth or in the neonatal period.

Eighteen of the 19 (95%) children with cervical CL (figure 1) presented within the first 2 years of life (age range 1 day to 7 years (median 6 months)). The size of the cysts ranges between 6 – 22cm in the longest axis (median 15cm). In two of the patients there was extensive involvement of the floor of the mouth. The right side of the neck was the most commonly involved in 57.9% of the cases seen. Preoperative complications were seen in 4 children with cervical CL (21.05%), two presenting with bleeding into the cyst, 1 presenting with stridor and infection respectively. None of the patients had retro-sternal or mediastinal extension.

Intra-abdominal abdominal CL presented much later in childhood with age range between 13 days to 13 years (median 7 years). 4 out of the 5 children presented after two years. Abdominal pain and swelling were the predominant symptoms at presentation. 2 of the patients presented with acute abdomen while 1 patient each had chronic abdominal pain and rapid increase in size of the mass respectively. Diagnosis was suspected preoperatively in 2 patients. In others the diagnosis was established at surgery and was confirmed by histopathology. The size of the cysts ranged between 6 x 6cm, cyst in the ileal mesentery to a massive cyst weighing 6kg in the omentum. These cases have been previously reported. [7]

In one of the patients with groin CL there was extension into the retroperitoneal space while in the child with axillary cyst there was involvement of the upper arm.

Treatment and outcome

Twenty four children had surgical excision. One patient with cervical CL died on the operating table as a result of asphyxiation that resulted from traumatic

rupture of a laryngeal cyst during intubations with subsequent drowning in the cyst fluid. This was confirmed at autopsy. There was failed intubation in another patient who was subsequently taken away from the hospital by the parents. The remaining 2 patients had no treatment.

Fifteen children with cervical CL had surgery. Complete excision was possible in 8 patients (53.3%) while in 6 (40%), gross complete excision was not possible because of attachment to surrounding vital structures but all visible cysts had their wall excised as much as possible. The remaining patient had partial excision as part of a planned staged surgery but was subsequently lost to follow up.

Six cysts were found in the 5 children with abdominal CL. Complete excision was possible in 5.. Bowel resection was necessary in 2 patients due to adhesion to the adjacent bowel. A child with an ileal mesenteric CL had partial wall excision and intra-peritoneal drainage of the cavity.

Complete excision was carried out in one patient with inguinal CL, while the another patient had partial excision. He later had three recurrences and repeated surgery. The axillary and breast CL were completely excised. Postoperative drainage was necessary in all cervical CL, axillary and breast cyst. Average duration of drainage was 7 days (range 2-21days).

Postoperative complications occurred in 10 patients (41.7%), 9 of whom had cervical CL. (table 2).

Follow up was 3 months – 7 years. One patient who had partial excision developed recurrence (4.6%).

Table 1: Age and site distribution of cystic lymphangioma in 28 patients

Site	Age (yrs)					Total (%)
	< 1mth	< 1	1-5	5-9	10+	
Cervical	8	7	3	1	-	19 (67.9)
Abdomen	1	-	1	1	2	5 (17.9)
Groin	-	1	1	-	-	2 (7.2)
Axilla	-	-	1	-	-	1 (3.5)
Breast	-	1	-	-	-	1 (3.5)
Total	9	9	6	2	2	28 (100)

Table 2: Postoperative complications in 24 children with cystic lymphangioma

Complications	No. (%)
Seroma collection	7 (29.6)
Wound infection	2 (8.3)
Facial nerve palsy	2 (8.3)
skin disfigurement	2 (8.3)
Inspiratory stridor	2 (8.3)
Hypoglossal nerve palsy	1 (4.2)
Recurrences	1 (4.2)

Figure 1: Left cervical cystic lymphangioma

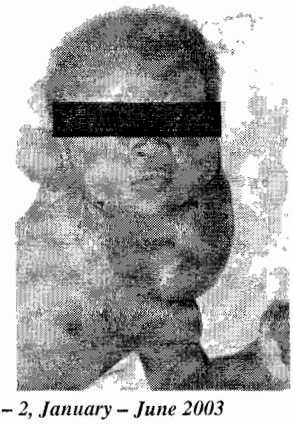
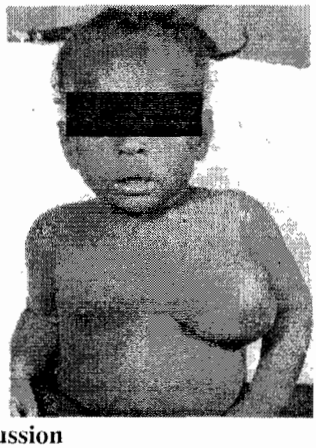


Figure 2: Lymphangioma of left breast



Discussion

The distribution of cystic lymphangioma is diverse but it occurs more commonly in the cervical region involving the posterior triangle of the neck. Whatever the location, the embryology is similar. [8] The pattern of presentation of the patients with lymphangioma in our own environment does not differ from what has been described in the literature. [9-12] The percentage of intra-abdominal CL in our series is however higher. However our patient population is small and most published articles tend to separate cervical cystic lymphangioma from others because of its unique features and management problems.

Preoperative morbidity is present in a significant proportion of patients presenting with cystic lymphangiomas. [9-12] This is also reflected in our patients' population. Overall 8 (28.6%) of our patients had complications at presentation. The incidence of preoperative morbidity was quite high in patients with intra abdominal CL. This may be because in patients with abdominal CL, presentation is often delayed and it is not unusual for the patient to present with pseudo-appendicitis or pseudo-ascites features as seen in two of our patients

The diagnosis of cystic lymphangioma is usually made clinically but plain radiographs, ultrasonography, computerized tomography (CT) scan and magnetic resonance (MR) scan where available, are used to confirm the diagnosis. These facilities are only becoming available recently in specialized centers in this country. It is believed that with improved education and access to health care, diagnosis would be made earlier and the incidence of preoperative morbidity would be reduced.

The management of cervical CL is difficult and operative excision and anesthesia is fraught with danger. Because of this, many new techniques of treatment have been developed in order to avert the morbidity and mortality that follows surgical excision. This includes the use of sclerosants such as OK 432, bleomycin emulsion, intra-lesional triamcinolone, and fibrin sealant with variable results. [3-6]. The use of OK 432 and other sclerosants are best suited for unilocular cysts and the patients need to be carefully selected. Surgery remains the gold standard against which all these new procedures will be compared and is the only resource available to surgeons in our environment. Complete surgical excision is the best where possible but operative excision of cervical cystic hygroma only becomes difficult and dangerous if complete excision is insisted upon because of the infiltrative nature of the lesion and attachment to important structures in the neck. Complete excision was possible in 8 out of the 15 (53.3%) children with cervical CL in our series compared to 41% from Zaria, Nigeria and 76% from Lagos Nigeria. [9,10]

One patient died because of anaesthetic complication. Although rarely reported, anesthetic death is an ever-present danger in the management of cervical CL. With the availability of CT scan, proper delineation of the anatomy of the airway and the relationship of the cysts to it will reduce this risk. Postoperative ventilation may be necessary where there is injury to the larynx or the recurrent laryngeal nerve, [14]

or following excision of very large cysts.

Patients with intra-abdominal CL are often misdiagnosed clinically and correct preoperative diagnosis is rarely made. [13] This is because of its rare nature and the absence of specific clinical manifestations. With the widespread use of the ultrasound and the computerized tomography diagnosis can now be made more easily preoperatively. [15, 16] This facility became available in our center only recently hence, in only 2 out of the 5 patients with intra-abdominal CL was the diagnosis made correctly preoperatively. Abdominal CL was easier to excise completely because they are usually related to the mesentery or the omentum. Bowel resection may be however be necessary if there is adhesion between the cyst and the adjacent bowel to allow for complete resection as in 2 of our patients.

Postoperative morbidity was quite high in this series. This is in agreement with findings elsewhere in Nigeria and other countries. [9-12] The commonest complication was postoperative seroma collection under the skin flaps. This can be explained by the fact that partial excision was done in a significant number of the patient leading continuous exudation of fluid from remnants of the wall left behind. This responded well to drainage and aspiration in all cases. All the complications resolved with conservative management.

In this series only one patient has reported recurrence but this is not likely to be a true reflection of the incidence since only 62.5% of those who had surgery had complete excision and it is known that incomplete excision increases the likelihood of recurrence. [2] Also many of the patients were lost to follow up after only a few clinic visits.

In conclusion, cystic lymphangioma in children have variable presentation and surgery remains the best and only option in this environment to treat these children but may need to be staged in order to avoid morbidity and mortality including injury to neighbouring structures. Complete excision should not be at the expense of anatomical and functionally important structures. There is however a significant pre and postoperative morbidity associated with these lesions especially with the cervical CL. Adequate attention should be paid to anesthesia in these patients

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