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Lymphangiomas in children

Ali AVANOĞLU, Volkan S. ERİKÇİ, İbrahim ULMAN, Geylani ÖZOK
Ege University Faculty of Medicine, Department of Pediatric Surgery, İzmir, Turkey

Summary

Fourty-one patients presented with a diagnosis of lymphangioma to the Department of Pediatric Surgery between 1971-1995 and 36 of them were treated. The distribution of lymphangiomas was: cervical (41.5 %), trunk (17.1 %), craniofacial (14.6 %), intraabdominal (7.3 %), axillar (7.3 %), extremity (4.9 %), cervicoaxillothoracic (2.4 %) and cervicoaxillar (2.4 %). Of 36 patients undergoing primary therapy, total excision was performed in 77.8 % and partial excision in 16.7 % with recurrence rates of 3.6 % and 83.3 %, respectively. Only two patients required drainage alone without clinical recurrence. Resection for residual or recurrent disease accounted 11.4 % (5/44) of all operations.

The postoperative complication rate was 29.5 %. Seroma was the leading complication with an incidence of 38.4 %. Neither drains nor antibiotics were found to be effective in preventing the occurrence of complications. The results of pathological examinations of this series were: cystic lymphangioma (55.9 %), simple lymphangioma (32.3 %), lymphohemangioma (8.8 %), cavernous lymphangioma (2.9 %). The overall mortality rate was 4.9 %. Only one patient was lost in the operative group due to pseudomonas sepsis. The mainstay of therapy for lymphangiomas is still complete resection of tumoral mass without compromising vital structures. It appears that local drains and perioperative antibiotics do not diminish postoperative complications.

Key words: *Lymphangioma, children*

Introduction

Lymphangiomas are benign tumors of the lymphatic system that appear to arise from congenital malformation of the lymphatics. Histologically they are benign tumors and malignant degeneration in unlikely. These asymptomatic, slowly growing lesions have a tendency of massive expansion and infiltration of surrounding adjacent tissues. This property makes total resection challenging and often incomplete. Thirty three to sixty seven percent of lymphangiomas are apparent at birth, and most of them appear by the end of the second year [2-4,6,10,11,13-15]. There are many series in the literature dealing with clinical characteristics, pathological findings and surgical efforts.

In this report it is attempted to define the management of lymphangiomas, their outcome and factors that might be related to morbidity and mortality.

Materials and Methods

A retrospective study was performed with the analysis of records of 41 lymphangioma cases presenting between 1971-1995 to our department. The patient data included age at diagnosis, sex, symptoms, signs, location, investigations, extent of each operative intervention, use of drains and antibiotics, pathology, complications and their treatment and outcome.

Results

Out of 41 cases twenty-two were male and 19 were female with a sex ratio of 1.2, and mean age was 3.3 years (Table I). The location of lymphangiomas was: cervical 17 (41.5 %), trunk 7 (17.1 %), craniofacial 6 (14.6 %), intraabdominal 3, axillar 3, extremity 2, cervicoaxillothoracic 1, cervicoaxillar 1 and multiple 1. Symptoms attributable to the lymphangioma were reported in two patients (4.8 %) at presentation and included symptoms secondary to local infection in one and vomiting and inability in swallowing in the other.

Physical signs of lymphangioma were reported in all the patients. An isolated mass was present in 70.7 % (29/41) and expanding in additional 4.9 % (2/41). Others included abdominal distention or abdominal

Address: Ali Avanoğlu MD, Ege University Faculty of Medicine, Department of Pediatric Surgery, 35100 Bornova, İzmir, Turkey

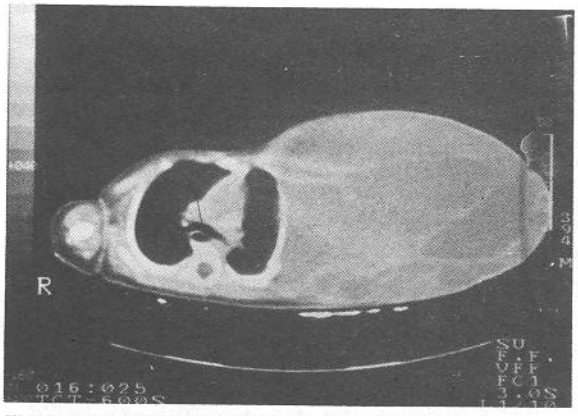

Figure 1. CT image of a giant lymphangioma case.

mass in 4, significant deformity such as lingual or cervical gigantism in 4, elepantiasis in one, cutaneous vesicles in one patient.

In addition to simple radiographs, ultrasonography, computerized tomography or magnetic resonance imaging were also performed in 11 (27 %) patients (Fig. 1) . Lymphoscintigraphy was performed in 2 patients and splenoportography in one. In our series there is only one patient who was diagnosed in the antenatal period by ultrasound. 36 out of 41 patients were operated on in our department by the same surgical team. Total and partial excision was performed in 28/36 (77.8 %) and in 6/36 (16.7 %), respectively. Only two patients required drainage alone without clinical recurrence (5.5 %). Recurrence rates for total and partial excision were found to be 3.6 % and 83.3 %, respectively.

A total of 44 operations were performed in 36 patients undergoing operation for primary and recurrent lymphangiomas. Resection for residual or recurrent disease accounted for 11.4 % (5/44) of all operations. The complication rate was 29.5 % (13/44). Distribution of these complications is depicted in Table II. Local drains were used in 25 operations (56.8 %) (9 hemovac, 16 penrose). The average duration of drain stay in the operation field for hemovac and penrose drains is 6 and 3.3 days, respectively. Antibiotics were given perioperatively in 20/36 patients (55.5 %). Other patients were handled without antibiotics. Complication rates in antibiotic and nonantibiotic group were found to be 25 % (5/20) and 18.7 % (3/16), respectively ($p>0.05$).

Table I. Age at diagnosis of lymphangioma

Age	n	%
Antenatal	1	2.4
Newborn	3	7.3
<1 yr	13	31.7
1 yr-5 yr	17	41.5
>5 yr	7	17.1
Total	41	100

Table II. Complications in 44 operations for lymphangiomas

Type	n
Seroma	5
Hematoma	1
Wound infection	1
Skin flap necrosis	1
Facial nerve palsy	1
Systemic infection	1
Chylothorax	1
Ptozis	1
Short bowel syndrome	1
Total	13

The results of pathological examinations of this series were: cystic lymphangioma 55.9 % (19/34) simple lymphangioma 32.3 (11/34), lymphohemangioma 8.8 % (3/34), cavernous lymphangioma 2.9 % (1/34). The overall mortality rate was 4.9 % (2/41). One patient died due to laryngeal pressure before the operation. The other patient with a complication of chylothorax died of pseudomonas sepsis in her postoperative 27th day.

Discussion

Congenital abnormalities of lymphatic system namely lymphangiomas are rare lesions cause of which are believed to be a developmental defect or primary malformation of lymphatic channels [12]. The true incidence is not known, and embryological events that lead to these malformations are still unclear [1-8,10,11,14,15]. It is reported that these lesions present at birth in 33 % to 67 % of cases [2-4,6,10,11,15]. In the present series one patient was diagnosed in the prenatal period and 51.2 % (21/41) were noted at birth.

The most commonly involved regions are head and neck with an incidence of 36 % to 88 % of lesions [2,3,5-7,10,13,14]. In the present series 60.9 % (25/41) cases were head and neck tumors including craniofacial, cervicoaxillar and cervicoaxillothoracic. The trunk followed by the abdomen, axilla and extremity tumors, were next most frequent sites. Majority of patients (70.7 %) presented with a simple mass and gradually expanding in additional 4.8 %, compared with 17 % in Brock's series [3]. It is reported that 16 % of cases present with local infection [13]. In the present series this occurred in 4.8 % of patients. Following clinical assessment, ultrasound, computerized tomography and magnetic resonance imaging are the most useful investigations for conforming the diagnosis and establishment the relationship to neighboring structures.

Although spontaneous regressions of these lesions have been reported to occur [11,13], this was not found in any of patients in the present series. Excision remains the treatment of choice once the diagnosis is established. Because, as the time passes, incidence of infections, hemorrhage and progressive growth that makes operative resection challenging increases. There are other methods of treatment including aspiration, incision and drainage, irradiation and chemical sclerosis. But the results of these treatment modalities have been unacceptable [1,3-6,10,11,13-15]. Aspiration may still have a role in emergency decompression but is not a definitive treatment. It may precede resection for massive lesions that cause respiratory obstruction. In our series one patient with facial and lingual lesions was handled emergently with aspiration. Elective surgical removal will be performed later on.

Recurrence rates range from 0 % to 27 % after total excision and up to 100 % following partial excision [3,7,11,14,15]. In the present series 77.7 % of lesions were totally excised whereas 16.6 % (6/36 were partially excised with recurrent rates of 3.6 % (1/28) and 83.3 % (5/6), respectively. Resections for residual or recurrent disease accounted for 11.4 % of all operations performed for lymphangiomas. Thus, morbidity in terms of additional operation is significant for partially excised cases in this series. Complete resection is the aim of surgical therapy. But it is known that these lesions are not malignant, so an extensive operation with sacrifice of vital structures is not warranted [2-4,14]. Recurrences generally occur within the first year following resection, in our series this period lasted as long as 10 years after the initial operation [7,11,14].

Complication rate following resection for lymphangiomas ranges from 19 % to 33 % [5,11,13]. In the present series this rate was found to be 29.5 %. The local wound and neurological complications accounted for 61.5 % and 15.3 %, respectively and were similar those reported. However, we also observed chylothorax and short bowel syndrome after surgical removal of the original tumor.

The use of drains and antibiotics at the time of operation has been addressed in some series. There is suggestion of usage of drains to prevent seroma formation by some authors [2,4]. Complications occurred in 48 % (12/25) of our cases in whom local drains were used. Antibiotics were given perioperatively in 55.5 % (20/36). Complications including those with infectious origin were seen in 25 % (5/20) of these operations as compared with 18.7 % (3/16) in which antibiotics were not used suggesting that usage of antibiotics do not diminish postoperative complications. However, the numbers are small and controlled studies are necessary to establish the statistical significance of these observations.

The overall mortality rate for lymphangiomas ranges from 3.4 % to 5.7 % [8]. In this series the overall mortality rate is 4.9 % (2/41). If nonoperative group is excluded, only one case in this series was lost due to pseudomonas sepsis in her late postoperative course.

Although it is reported that intralesional therapy with bleomycin and OK-432 may be an alternative therapy, resection still remains the mainstay of therapy [9]. Complete excision without compromising the vital structures is the gold standard of therapy for lymphangiomas. The benefits of drains and perioperative antibiotics are unclear, because they do not appear to diminish postoperative complications.

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