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Cystic hygroma presenting at an unusual site – A case report

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Abstract

Cystic hygroma (Lymphangioma) is a benign congenital cystic malformation that occurs mostly in children. Their incidence in paediatric population rises up to 1 new case per 12000 births, while lymphangiomas represent almost 5-6% of all tumors in childhood. It is commonly located in posterior triangle of neck, axilla, face, cervix, groin and oral cavity. It can occur anywhere in the body and is rarely found in abdominal wall and chest wall. Lesions from chest wall can extend into mediastinum. The unusual site of a cystic lymphangioma in the abdominal wall was the real motive for the present case report. We report a case of a 14-year-old male presenting with a large swelling in hypogastric region, involving bilateral inguinal region and extending into the scrotal sac since birth. Two small swellings in right hypochondriac and umbilical region were also present since birth.

Keywords: Abdominal, Cystic hygroma.

Introduction

Cystic Hygroma is also known as Cystic lymphangioma. It is a benign congenital malformation characterized by proliferation of lymphatic vessels, resulting from the failure of communication between the primitive lymphatic sacs and the venous system, leading to the formation of a cystic structure [1]. Rarely, it is an acquired malformation, due to inflammation, injury, or fibrosis [2]. Their incidence in paediatric population rises up to 1 new case per 12000 births, while lymphangiomas represent almost 5-6% of all tumors in childhood. Almost 95% of all lymphangiomas are detected in the neck, face and axillary region [3]. Other sites are in the maxilla, superior mediastinum, mesentery, retroperitoneal region, pelvis and lower limbs [4,5]. Rarely, it occurs on the abdominal wall. There is no predilection for either sex [6]. Based upon the histopathologic features, lymphangiomas are divided into three subtypes: the capillary (macrocystic)

lymphangioma, cystic lymphangioma (Supermacrocystic) and the corpus (cavernous, microcystic) lymphangioma, which is the rarest [7]. As for aetiology, according to embryologic mechanism, septate lymphangioma results from total obstruction of the primitive lymphatic sacs, while non-septate lymphangioma from partial obstruction [8]. In our case parents noticed the swelling on abdominal wall and bilateral descended testis, which was present since birth. CECT findings of abdomen and pelvis were suggestive of veno-lymphatic malformation of abdominal wall. Histopathological examination of excised lesion confirmed the clinical diagnosis of cystic lymphangioma.

Case Report

14 year old male presented in surgical OPD with swelling over lower abdomen extending into bilateral scrotal region (11 x 11 cm). Two small swellings located 2cm right from the umbilicus and below the right rib respectively were present. A reddish patch was noted on back and anterior side of chest extending into abdomen since birth.

On local examination, a mass of size 11 x 11 cm was present over lower abdomen involving hypogastric, and bilateral inguinal regions extending into bilateral scrotal region. The mass was soft to firm, non-tender and mobile. (Figure1). Two smaller masses measuring 3 x 3 cm and 2 x 2 cm and of similar consistency were noted in the right hypochondriac region and umbilical regions respectively. Anterior and posterior side of chest wall showed well defined hyperpigmented patches with overlying hyperpigmented (brown to black) papules of size 12 x 7cm and 9 x 8 cm respectively.

CECT Scan Abdomen and Pelvis showed a large well defined predominantly cystic multi septate lesion measuring 11.8 x 13.4 x19.3 cm in lower anterior abdominal wall in hypogastric region, which extended into bilateral inguinal scrotal region. Right anterior and lateral abdominal wall upto thoracic region appeared to be thickened with multiple lymphatic channels within. Bilateral testes were not visualized in respective scrotal sac. Right testis msg 2.1 x 1.8 cm was noted in Rt. inguinal region. Left testis msg 1.8 x 1.5 cm was noted in left inguinal region. CECT findings of abdomen and pelvis were suggestive of veno-lymphatic malformation of abdominal wall. HRCT thorax Right anterior and lateral thoracic wall appeared to be thickened with multiple lymphatic channels within it.

Clinical diagnosis of Cystic hygroma with Bilateral undescended testis was made. Multiloculated cystic swellings from bilateral scrotum was excised and sent for histopathological examination. Injection bleomycin was injected and repositioning of testis was done.

On gross examination we received an irregularly shaped tumor mass, firm in consistency, measuring 18.5 x 12 x 5.5 cm. On cutting open greyish white areas, few fatty areas and cystic spaces were seen. The largest cystic space measured 4 x 2 x 2 cm (Figures 2 and 3).

Microscopic examination: Multiple sections from different areas of the specimen were studied. They revealed many variably sized cystic spaces lined by flattened endothelial cells. Their lumen contained eosinophilic material with few scattered lymphocytes. Intervening fibroconnective and adipose tissue with scattered and aggregates of lymphocytes were seen (Figure 4). Many congested, dilated blood vessels and smooth muscle bundles surrounding the cystic spaces were present

The histopathological diagnosis was Cystic hygroma.

Discussion

Cystic hygromas/lymphangiomas are thought to be developmental abnormalities associated with a failure in the embryological connection between lymphoid vessels and venous system and are generally not accepted as true tumors [9]. The lymphatic system develops as a sac -like out growth from the endothelium of veins in the fifth week of gestation. There are six primary lymph sacs of which two develop in jugulars, two in iliac, one in retroperitoneal and one in cisterna chyli respectively. Failure of communication with lymph system results in formation of cystic lymphoma. Majority of cases present in the first 2 years of life and 65% are noted at birth. Pan S. et al in 1843 described the various sites of Cystic lymphoma [10]. Goldstein et al. focused on the prenatal diagnosis of cystic lymphangioma during the 15th and 22nd week of gestation by USG during the nuchal translucency test. About 75% cases are in the neck often in posterior triangle, 20% in axilla and the remaining 5% in other parts of body [11]. Although it can occur in any anatomical site in human body, the head and neck is affected more, with a predilection for the left site. The other affected sites are the mediastinum, groin and retro peritoneum. Rare sites of occurrence are anogenital region, forehead, orbit, buttock, tongue, gastrointestinal tract, extremity and pelvis [10]. Hancock et al reported various sites of involvement of cystic lymphangioma as cervical-31.45%, craniofacial-18.9%, extremities-18.9%, trunk-9.2%, intraabdominal-9.2%, cervicoaxillothoracic-4.9%, multiple-3.8%, cervicomediastinal-2.25% and intrathoracic-1.6% [11].

Pandit et al reported two cases of Cystic lymphangioma in the abdominal wall (4.4%) [4]. Very few cases of Cystic lymphangioma of abdominal wall are published so far [12,13]. Kyeom Kim et al from Korea reported two cases of Cystic lymphangioma - one was in the abdominal wall in lower left quadrant and in the other case was in the right upper quadrant of the abdominal wall [11]. Ammar et al from University of Tunisia reported a case of cystic lymphangioma in a 10 years female child, in the left side of abdomen, with mass extending to pelvic cavity [12].

Aspiration of the lesion may result in reduction in size temporarily only. Surgical excision of the lesion is considered as a standard treatment. Incomplete excision often results in recurrence. Other surgical complications are damage to surrounding blood vessels, nerves, hypertrophied scar and lymphatic discharge from the wound [14].

Three types of cystic hygroma/lymphangioma can be distinguished. The capillary form is usually asymptomatic with small sizes. Although cavernous and cystic lymphangiomas show the same histological pattern, cystic lesions are usually larger and symptomatic [15]. Microscopically, lymphangioma is characterized by large, dilated lymphatic vessels in a fibrotic or loose stromal background. Focal areas of papillary endothelial proliferation were described aside by the classic histological picture [16]. The presence of lymphoid aggregates even lymphoid follicles may be confusing when existing in large amounts and needs to be distinguished from atypical lymphoid proliferations. The main histopathologic differential diagnosis of cystic hygroma is cavernous haemangioma in which blood filled large cystic spaces are seen, similar to lymphangioma. The lining endothelium of cavernous haemangioma shows positivity with pancytokeratin and Factor VIII, immunohistochemically. The presence of lymphatic spaces with thin walls containing fibrous

tissue, smooth muscle, and lymphoid aggregates favours the diagnosis of lymphangioma [17].

In general, lymphangioma is accepted as a benign tumor with no malignant transformation and curable by excision [18]. If a cystic lymphangioma is fully excised, possibility of relapse is rare [19]. Infiltration of the adipose tissue constitutes a negative prognostic factor, increasing the possibility of relapse after excision [20]. In cases of incomplete removal, relapse should be expected within the first 3 months postoperatively. Flanagan et al. reported a case of relapse 7 years postoperatively, while Lee et al. report a case of a chest wall cystic lymphangioma that relapsed 19 years after surgical removal [21].

Cystic lymphangiomas are amongst the vascular anomalies associated with Maffucci and Cobbs syndrome, Turners syndrome, diverse congenital malformations, several chromosomal aneuploidies and foetal death [22].

Conclusion

In our case, cystic lymphangioma presented in the abdominal wall, which is a unusual site.

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Legend Figures

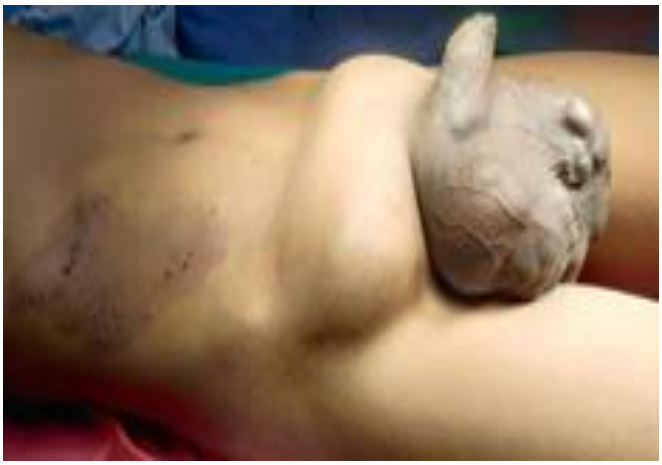

Figure 1: Clinical photograph showing mass in lower abdominal wall extending into the scrotal sac

Figure 2: External surface of excised specimen was greyish brown and bosselated.

Figure 3: Cut surface of excised specimen showing many cystic spaces.

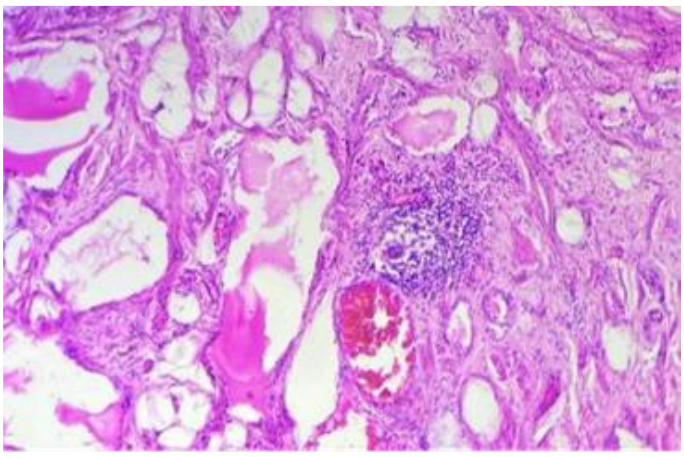

Figure 4: Photomicrograph showing many cystic spaces filled with eosinophilic proteinaceous material and surrounding lymphoid aggregates (Hematoxylin & Eosin stain 10x)