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Lymphangioma of the trunk presenting as inguinal hernia: An unusual presentation

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Abstract

Lymphangiomas are malformations of the lymphatic system. They are abnormal proliferation of the lymph vessels producing fluid field cyst lined by endothelial cells. These result in sequestrations of lymphatic tissue that do not communicate with the normal lymphatic system. A small proportion appears to arise from localised lymphatic malformations or obstruction.

Lymphangiomas are rare, account for 4% of all vascular tumors and approximately 25% of all benign vascular tumors in children.

Chromosomal abnormalities are usually associated with other congenital findings such as mental or developmental retardation, heart defects, or alterations in the development of sexual characteristics at puberty.

The patient was managed by excision of the lymphangioma and did well with little residual of the trunk mass. Surgical resection should follow the precise documentation of the lesion with MRI or CT scan and should remove the entire lesion whenever feasible.

Keywords

lymphangioma; congenital; malformation; chromosomalidney; renal malformations; infancy

Introduction

Lymphangiomas are malformations of the lymphatic system. They are abnormal proliferation of the lymph vessels producing fluid field cyst lined by endothelial cells that result from maldevelopment or blockage in the lymphatic [1,2]. This result in sequestrations of lymphatic tissue and these do not communicate with the normal lymphatic system.

Lymphangiomas are rare. They account for 4% of all vascular tumors and approximately 25% of all benign vascular tumors in children [3]. In most part of Africa, incidence data are not available, but hospital-based reports suggested that at least 1–3 children with lymphangiomas are seen every year in most teaching hospitals. Lymphangiomas are benign but frequently presents surgical difficulties and challenges due to their propensity to infiltrate and extend around neighbouring structures [4,5].

They are most commonly located in the head and neck region, and to a lesser extent on the axilla and trunk, but can occur anywhere there are lymphatic vessels. Even though they are congenital defects,

they may not become apparent until several months or years after birth [6]. Lymphangioma involving the trunk or retroperitoneum are rare. We herein present a 2 year old boy with lymphangioma of the trunk extending in to the abdominal cavity, chest and scrotum.

Case Report

IA is a 1yr old male, a product of full term gestation. He presented with a Swelling involving the lateral aspect of chest and abdominal wall noticed at birth, which grew slowly with no history of ulceration, bleeding or discharge over the swelling. There was no history refusal to feeds, vomiting, abdominal distension, constipation or weight loss. There was no history of cough, or difficulty in breathing. His Pregnancy was booked at 5months gestational age and his mother took routine antenatal drugs. His Labor and delivery were at home via spontaneous vertex delivery. There was no maternal illness during pregnancy, no ingestion of herbal or over the counter medications during pregnancy, No undue exposure to radiation. His father is a 25years old trader and the mother a 20yr old house wife. On examination he was well preserved child, not in distress, afebrile, pale, not dehydrated with no pedal edema.

Respiratory rate was 34 cycle per minute, with vesicular breathe sounds. His heart rate was 120 beat per minute, heart sound 1 and 2 only, no murmur. He had a huge swelling on left side of the trunk extending from axilla to the iliac crest. It extends anteriorly to just before the midline and posteriorly to the infrascapular region. The mass was Irregular in shape, measuring 30x24cm, with no differential warmth. It was nontender and attached to the overlying skin, with hyperpigmentation and nodules. He had an umbilical swelling which was soft, nontender, and reducible with fascial defect of 1.5cm diameter. There was a left Inguinoscrotal swelling which was soft, nontender and not reducible. There was an enlarged 1st and 2nd toes with intact skin 4x4 and 8x6cm respectively.

Figure 1: Lymphangioma of the trunk.

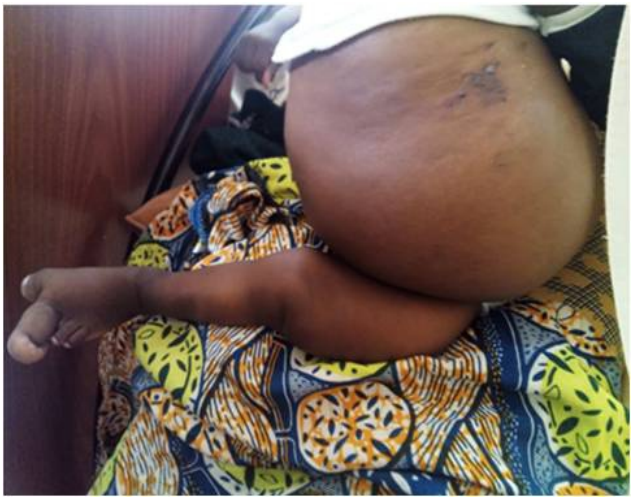

Figure 2: Lymphangioma of the trunk with involvement of the second toe

Thoraco-abdominal CT showed a Heterogenous soft tissue mass over the left side of the trunk displacing left kidney and bowel to the contra lateral side. The mass was also extending via inguinal canal into the scrotum (Figure 3).

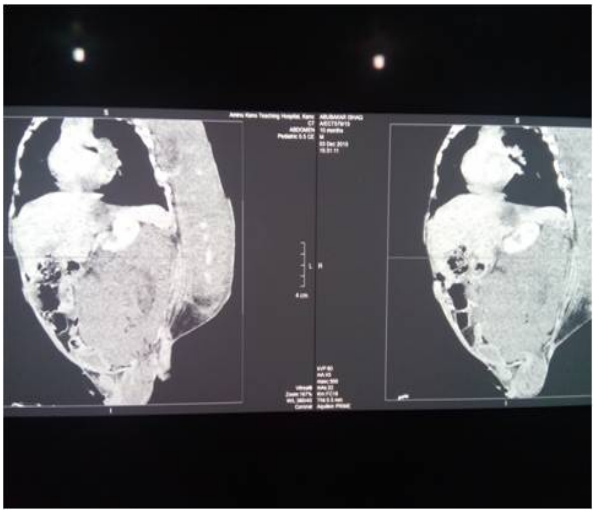

Figure 3: Thoraco abdominal CT

His full blood count showed haemogram of 12.5g/dl, 2 units of blood was grouped and cross matched and an informed consent was obtained in preparation for the surgery.

The surgery was done under general anesthesia with endotracheal intubation, in a supine position. Intra-operatively a Multilobulated cystic mass in the left thoracoabdominal wall was seen extending in to the retroperitoneal space and left inguinal region pushing the left kidney and bowel to the right side. A small part of the tumor was seen extending in to the chest via the esophageal hiatus.

A debulking excision was done, the masses on the trunk, retroperitoneal and left scrotum were excised, but the chest extension in the posterior mediastenum and the left second toe were left untouched. 2 corrugated rubber drains were inserted in the retroperitoneal and left trunk, and Specimen sent for histology which revealed CAVERNOUS LYMPHANGIOMA.

The mass on the left second toe was excised by the plastic team about a month later and patient followed up in the out -patient clinic.

Patient did well post op, but lost follow up about 4month after the surgery.

Discussion

The lymphatic system starts to develop by the end of the 5th week of gestation, 2 weeks after the primordia of the cardiovascular system are recognisable and 1 week after coordinated contractions of the primitive heart initiate unidirectional blood flow. The lymphatic vessels are thought to be derived from the venous system as endothelial out growths. By the 8th week of gestation, six lymphatic sacs are formed: two jugulars, two iliac, the retroperitoneal sac at the root of the mesentery, and the cysterna chyli [4]. These sac developed in to lymphatic vessels that drained the excess tissue fluid back in to the vascular circulation. The majority of lymphangiomas arise from parts of lymph sacs that are pinched off during development or that fail to establish connections with the main lymphatic or venous channels.

Lymphangiomas are rare. They account for about 4% of all vascular tumors and approximately 25% of benign vascular tumors in children. Lymphangioma affecting the retroperitoneal region and trunk account for about 3% to 9% of all lymphangiomas [6]. The aetiology of lymphangiomas is unknown, however, cystic hygromas have been found to be associated with chromosomal abnormalities such as Turner syndrome and Down syndrome [7].

The age at diagnosis ranged from prenatal period to few years after birth. Rarely, it can present in adolescence or adulthood. Our patient presented at the age of 1 year but the swelling was noticed immediately after delivery, which is in keeping with most studies on lymphangioma [8]. Prenatal diagnosis even though rare in sub Saharan Africa is increasing worldwide, in our index patient the pregnancy was booked at 5 month gestation, mother had ultrasound scan once at 7 month gestation but could not detect the anomaly prenatally. Diagnosis is principally made on the basis of clinical appearance and imaging [8,9]. In our patient the diagnosis was made based on history and examination and CT was able to show the intra thoracic extension.

Lymphangiomas are benign vascular malformations; therefore, growth is not a concern, although the size of the lesions varies over time [10]. Small superficial lymphangiomas can be observed over time. The treatment options are surgical excision, scleratherapy, and radiotherapy [11,12].

Percutaneous sclerotherapy is performed with a sterile technique, usually under sedation or general anaesthesia, depending on the age of the child. With Ultrasound guidance, fluid is aspirated and sent for cell count and cytological examination to confirm the diagnosis. The sclerosing material is then introduced into the cysts [4,13,14]. Many sclerosing agents have been used, including absolute ethanol (98%), doxycycline, OK-432, and bleomycin.

Our Patient had surgical excision of the tumor with little residual tumor and the parent were satisfied with post operative appearance of the child.

Complications following treatment of lymphangiomas depend on the size, site and the methods of treatment. The two major complications of lymphangioma are intralesional bleeding and infection, which may occur prior to treatment or after the treatment. Other common complications are airway obstruction, damage to the Nerves, recurrence [14,15].

Our patient had surgical site infection which was treated before discharge from the hospital, the follow up period in our patient was short because parent lost follow up about 4 month after the surgery.

Conclusion

Management of lymphangioma in our environment can be challenging due to delay diagnosis, partly due lack of an established prenatal diagnosis, delayed presentation and in adequate facility and manpower to manage lymphangiomas.

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