CT in the Assessment of Paediatric Lymphatic Malformations

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ABSTRACT

BACKGROUND

Lymphatic malformations are a form of congenital vascular malformations. They are usually seen in the neck and axillary region, while mediastinal, abdominal, mesenteric and retroperitoneal locations are very rare and account for about 5% of cases. Imaging has a fundamental role in the detection and characterisation of these lesions. We wanted to evaluate the role of Computed Tomography in the assessment of paediatric lymphatic malformations.

METHODS

In our study, during a period of 1 yr. 7 months (October 2018 to April 2020) 30 children with the possibility of lymphatic malformations on ultrasound evaluation were subjected to CT examination. Various imaging features of lymphatic malformations on CT were noted.

RESULTS

30 paediatric patients, aged between 5 days to 8 years were included in the study. The most common age of presentation was less than one year age group. The most common site for the lymphatic malformations was neck region. One case had complication of haemorrhage and one case had secondary infection. Seven cases out of 30 showed enhancement of the thin internal septations. CT could delineate clearly the extent of the malformation in all the cases.

CONCLUSIONS

Accurate anatomic localization and definition of the lesions are important in preoperative planning because lymphatic malformations have an insinuating nature that makes complete surgical excision difficult in some cases. The diagnostic challenge on imaging is the differentiation of lymphatic malformations from other fluid-containing masses. CT is a useful imaging tool in confirming the diagnosis, and anatomical delineation for preoperative localisation of lymphatic malformations. It also helps in detecting associated complications like haemorrhage and secondary infection.

KEYWORDS

Lymphatic Malformation, Paediatric, Computed Tomography

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BACKGROUND

Lymphatic malformations are a form of congenital vascular malformations.[1] They are preferentially localized in the neck and axillary region while mediastinal and abdominal, both mesenterial and retroperitoneal, locations are very rare approaching 5% of cases.[2] Histopathologically, lymphatic malformations are characterized by thin-walled unilocular or multilocular cysts that are lined by endothelial cells and contain clear or milky fluid. The wall contains varying amounts of connective tissue, small lymphatic spaces and smooth muscle fibers.[3,4] Generally, lymphatic malformations present clinically as soft tissue masses. In some cases deeper lesions may not present as a palpable mass unless large in size. They may present due to pressure effect on adjacent structures. They do not respect tissue planes and insinuate into different anatomic planes and areas in continuity. They are considered to have no malignant potential.[5] Lymphatic VMs never involute, but expand or contract depending on the flow of lymphatic fluid, and the occurrence of inflammation or intralesional bleeding. Indications for treatment depend on the size, location and symptoms of the lesion.[6] Cosmetic disability, presence of recurrent infection, oozing, crusting, ulceration and pain are the most frequent indications for treatment.[7] Rarely serious functional disability may be seen when present in the orbit or larynx. Although surgical excision has been considered to be the treatment of choice by most of the surgeons, sclerotherapy of lymphangioma has gained popularity during recent years.[8] Imaging has an important role in management of the lymphatic malformations both in pre-operative planning and follow up. Radiologists should be familiar with the imaging spectrum of these lesions and with some common mimickers in the paediatric population so that prompt and accurate diagnosis can be achieved, and optimal patient care can be provided.[9]

Objectives

- To evaluate the role of Computed Tomography in the assessment of paediatric lymphatic malformations.
- To confirm the diagnosis or suggest an alternate diagnosis and to assess the anatomical extent of the lesion based on CT morphology.
- To assess structural features of the lymphatic malformations which help the clinician for planning the management.

METHODS

In this study, after informed consent, 30 paediatric patients admitted in Nilofer hospital for women and children, Hyderabad, with soft tissue mass and diagnosed with lymphatic malformation on ultrasound were evaluated with CT for a duration of one year seven months (October 2018 to April 2020). Our study included patients of age groups 5 days of life to 8 years with an equal proportion of female and male children. Patients were initially evaluated with routine investigations and later ultrasound examination. Ultrasound findings of uni or multilocular cystic lesions with thin septations with focal area of involvement or extending into multiple compartments were given a possibility of lymphatic malformations. Colour Doppler evaluation in the lesions showed no vascularity with in. These lesions were further evaluated with computed tomography.

CT examination was performed on Toshiba Aquilon 16 slice machine before and after intravenous contrast administration of Iomeprol which is a non-ionic water soluble contrast medium. The paediatric patients were put to sleep by the attendant naturally or a dose of Pedicloryl was given orally and the CT done after the child is calm. The lymphatic malformations were examined in axial, coronal and sagittal planes. On CT examination, the malformations were thoroughly examined to localise the lesion and confirm the nature of the lesion, either completely cystic or with solid areas. The size, extent of the lesion and its relation to the adjacent structures noted. Presence of internal septations, enhancement pattern of lesions and associated complications like haemorrhage and secondary infection noted. Associated findings of compression collapse of adjacent lung in thoracic malformations and displacement of bowel and solid viscera in abdominal malformations were noted in detail. Note of all the normal structures and any other pathology in the anatomical area scanned were also noted.

RESULTS

Age Group of Patient	No. of Male Patients	No. of Female Patients
Less than 1 yr.	4	8
1 to 5 yrs.	7	4
More than 5 yrs.	4	3
Total No. of Patients	15	15

Table 1. Distribution of Study Subjects According to Age

Table-1 out of the thirty patients, the most common age group was less than one year .next common age group was one to five years age group. There was an equal sex predilection in our study.

Location of Lymphatic Malformation	Number of Cases
Neck including pharynx and oral cavity	11
Neck and extending into thorax	3
Neck and extending into axilla and thorax	4
thorax	1
Intra-peritoneal	3
retroperitoneal	3
Soft tissue planes of abdomen, thorax with no extension into deeper planes	4
Gluteal and sacral region	1

Table 2. Different Sites of Lymphatic Malformations

Table-2 out of the thirty patients, the most common site of lymphatic malformation was in the neck region with eleven cases confined only to neck region and an additional seven cases of neck extending into thorax and /or axilla.

Next common site was subcutaneous and superficial planes of chest and abdomen.

Findings on CT	Number of Cases
Haemorrhage	1
Infection	1

Table 3. Findings and Complications Diagnosed on CT

Table 3, out of the thirty cases only one case had haemorrhage and another case had findings of secondary infection.

Figure 1

Figure 1. Contrast enhanced (a)coronal, (b)sagittal and (c) axial CT sections at the level of thorax show Cystic lesion with thin internal septations in superior mediastinum on left side extending into neck superiorly causing splaying of left subclavian and common carotid arteries (black arrow) and stretching of left brachiocephalic vein and also minimal compression collapse of adjacent lung parenchyma (*).

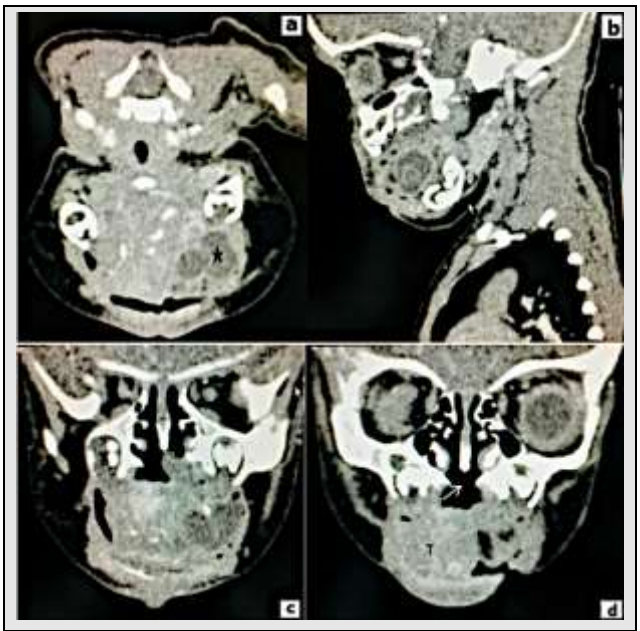

Figure 2

Figure 2. Contrast enhanced (a) Axial, (b) Sagittal and (c & d)coronal sections at the level of floor of mouth show Cystic lesion with thin enhancing internal septations in oral cavity (*) on left side extending up to nasopharynx displacing the tongue to right (T). There is associated cleft palate (white arrow).

DISCUSSION

The origin of lymphatic malformations is considered to be congenital abnormality of lymphatic system rather than true neoplasm.[10] The fact that most lymphatic malformations manifest clinically during early childhood and develop in areas where the primitive lymph sacs occur (neck, axilla) provides presumptive evidence for this hypothesis.[11] The other locations include the orbit, mediastinum, retroperitoneum, chest, abdomen, extremities, scrotum and penis.

Three subtypes based on morphologic features have been described: macrocystic, micro cystic, and combined. The macrocystic subtype is composed of locules measuring greater than 1 cm, the microcytic subtype comprises smaller (usually subcentimeter) cysts, and the combined subtype shares features of both.[12]

Symptoms depend on the location of the lesion.[13] Sub cutaneous lesions presented with palpable mass. The lesions with intra thoracic extension presented with associated chest symptoms of respiratory distress. The abdominal lesions presented with vague pain abdomen. Soft tissue lesions may be associated with overgrowth of adjacent skeletal structures, especially in cranio facial region. The abdominal location is rather rare and accounts for approximately 5% of all lymphatic malformation. Abdominal lymphatic malformation can arise from mesentery, omentum, gastrointestinal tract, and retroperitoneum.[1] An important feature on imaging that differentiates these lesions from other pathologies is the insinuating nature crossing multiple compartments.

Imaging can help in differentiating from other cystic lesions like cystic teratoma, haematoma, necrotic tumours and abscesses. Ultrasound is first line imaging modality due to its availability, non-invasiveness and cost benefit. It has the limitations of unclear lesion delineation and also not accurate about the structural details like enhancement pattern. Due to the panoramic view, Cross sectional imaging has the advantage of clear visualisation of extent of lesion .Contrast enhanced CT has also the advantage of lesion enhancement. As compared to magnetic resonance imaging examination, the shorter acquisition times and cost effectiveness of CT makes it useful for paediatric population. CT provides important information regarding lesion location, size, and shape; the presence and thickness of a wall; the presence of septa, calcifications, or fat and involvement of adjacent structures. However, CT imaging is performed with radiation exposure. Conversely, MR can give specific information about the fluid content even when the CT shows greater intralesional attenuation values for the presence of proteinaceous material.[14]

Our study of CT evaluation of lymphatic malformations consisted of 30 paediatric patients, of which 15 were males and 15 were females. Age of the patients varied from 5 days of life to 8 yrs. Commonest age group in our study was less than one year. As per literature, in 50%-60% of patients, these malformations are present at birth; 80%-90% of the lesions are detected by the age of 2 years.[5]

Most common site involved in our study was neck region with eleven cases confined to the neck, involving the deeper neck spaces and an additional seven cases have shown extension into the chest or / and axilla. Lymphatic

malformations can be of varying in size, as a small sub cutaneous lesion to large multiseptated cystic lesions extending in to multiple compartments. There was one case of lymphatic malformation in oral cavity extending into nasopharynx which had associated cleft palate picked up on the CT. The lymphatic malformations of the neck had extension either into axilla or into the thorax or both and these pose difficulty in surgical management. Due to insinuation of the lesions into various tissue planes, complete surgical removal is challenging, and recurrences do occur with such lesions. Few of the neck lesions appeared confined only to the neck extending into the pharyngeal soft tissues.

There were 4 cases which involved only the soft tissues of chest wall and abdominal wall with no extension into deeper planes. These can be completely excised with very less chances of recurrence. There were three cases of retroperitoneal lymphatic malformations which is considered as a rare site of the malformation. An important feature on imaging that differentiates these lesions from other cystic retroperitoneal lesions is the insinuating behaviour of crossing multiple compartments. The recurrence rate in abdominal lymphatic malformations depends on the lesion location, size, and complexity. One case had the lymphatic malformation in the gluteal soft tissues extending into perirectal space. The child had visible swelling with redness and pain in the gluteal region. CT showed enhancement of the thick walls and septations, in the gluteal region suggesting possibility of infection.

The most common CT appearance in our patients, a uniformly cystic mass, which is also the most common CT appearance reported in other series.[15] Seven cases out of 30 showed thin enhancing septations. There was one case of retroperitoneal lymphatic malformation with hyper dense areas with HU values of blood, suggesting possibility of haemorrhage. However hyperdensity of the contents in a lymphatic malformation can be due to proteinaceous contents. The delineation of the extent was clear on CT in all the cases. The pressure effects of some of the lesions on adjacent structures were also made out on CT. CT was also useful to diagnose additional findings like collapse and consolidation in the adjacent lung parenchyma in cases of thoracic lymphatic malformations. Three cases of lymphatic malformations of the neck extending into thorax had the above findings of collapse and consolidation.

The mainstay of management is to eradicate the lesion whilst causing minimal damage to adjacent structures. Various treatment modalities such as surgical excision, laser therapy, sclerotherapy or combination therapy have been proposed in management of such lesions.[16] Sclerotherapy is recently attracting attention and the procedure consists of transcutaneously injecting a sclerosant into the vessel, causing inflammation of the vessel wall, followed by fibrosis, with obliteration of the lumen of the vessel.

CONCLUSIONS

In paediatric population, CT is an efficient modality for the evaluation of lymphatic malformations. The shorter acquisition time and the panoramic ability of CT are useful. CT has the advantage of accurate delineation of the lesion and also provides better structural detail compared to ultrasound which helps in management of these lesions. CT also helps in the diagnosis of complications of lymphatic malformations like haemorrhage and infection. Radiologists should be well acquainted with the imaging morphology of these lesions in the paediatric population so that definitive diagnosis can be achieved which in turn helps in optimal patient management.

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