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CYSTIC LYMPHANGIOMA OF ABDOMINAL WALL IN AN ADULT, RARE SITE OF OCCURRENCE - A CASE REPORT.

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Abstract:

Cystic lymphangiomas are rare, congenital, benign lesions occurring early in life, mainly in the head, neck, and oral cavity and rarely in abdominal wall, Due to aberrant proliferation of lymphatic vessels resulting from abnormal development of the lymphatic system. Lymphangiomas are classified as micro cystic, macro cystic, and cystic hygromas according to the size of the lymphatic cavities incorporated. These are soft, variable in size and shape, and tend to grow extensively if not surgically excised. These are multilocular cysts filled with clear or yellow lymph fluid. Histopathologically, lymphangiomas are of three types: Lymphangioma simplex, Cavernous Lymphangioma, and Cystic Hygroma depending on the size of vascular spaces and thickness of the adventitia. The present case report describes a case of cystic lymphangioma of lower abdominal wall in a 30-year-old male and its clinical, ultrasonographic Imaging features, cytological and histo pathological correlation.

Keywords: Cystic Lymphangioma CL, Cystic Hygroma CH, Macro Cystic lymphatic malformation Abdominal Wall. Abnormal Abdominal Swelling AAS.

Introduction:

Cystic lymphangioma or Cystic hygroma is a congenital malformation due to failure of communication between the primary lymphatic sacs to drain into the venous system[1]. This results into the formation of a cystic structure[2]. Also known as macro cystic lymphatic malformation and was first described in 1828 by Redenbacker.CL commonly occur in neck, more than 80%.Other sites are in the maxilla, superior mediastinum, mesentery, retroperitoneal region, pelvis and lower limbs[4,5]Rarely it occurs on the abdominal wall. Few case has been reported in the literaure.[3,4,7,8,14]. Cystic lymphangiomas are slow growing tumors and usually located to one organ but once their infiltration into local tissue, surround muscles and adjacent nerve and vessels then surgical removal becomes difficult. In our case patient noticed the progress of the swelling on right lower abdominal wall with extension to right inguinal region.

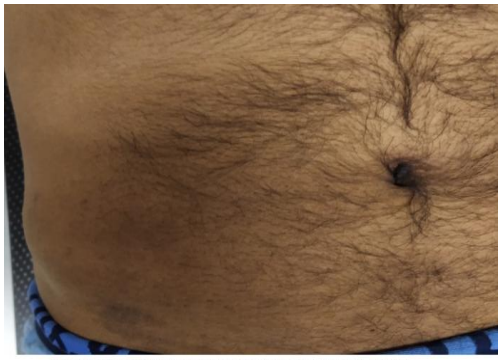
CASE REPORT

A 30 years old male was referred to the Radiology department for Radiograph of chest, abdomen and Ultrasound examination of swelling in Right side of lower abdominal wall. This swelling was progressively increased in size since last two years. On physical examination a palpable soft non tender swelling of 5x5 cm size was seen in subcutaneous plain of right lower abdominal wall and extending up to right inguinal region **(Figure -1-2).**The overlying skin was normal**.** The patient was in good health with normal developmental milestones. No other physical deformity or abnormality was seen in rest of the body.

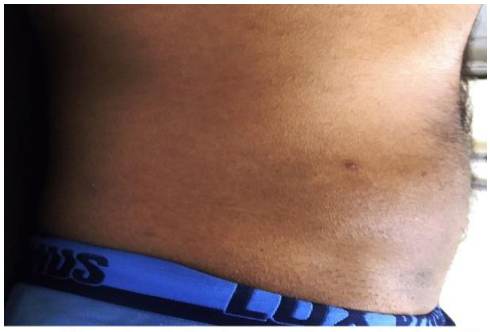
Chest Radiograph reveals no significant abnormality. A soft tissue swelling/ bulge were over right lower abdominal wall on skiagram of abdomen. No calcification was seen. **(Figure 3-4**).

Ultrasound study reveals subcutaneous multilocular clusters of abnormal cystic channels with internal echoes in right lower abdominal wall with inferiorly extension in right inguinal region. In few channels, thin septae were seen. There was no abnormal vascularity seen on color Doppler. No obvious invasion of muscles seen **(Figure 5-7**). Ultrasound of abdomen and pelvis reveals no abnormality. CT lower abdomen and pelvis findings showed multiloculated fluid density lesion of 2.3 x 12.2 x 13.4 Cms size in right lower abdominal wall in subcutaneous plane with extension up to right inguinal region. Few locales were hyperdense on CT images. There was no evidence of intra abdominal extension**.(Figure 8-11)**The aspiration cytology from the lesion showed predominantly lymphocytes against a proteinaceous background. Under all aseptic precaution and condition, this lesion was surgically excised with complete excision and preserving cosmetic function. The swelling showed cystic areas containing lymphatic fluid mixed with blood**. (Figure-12).**Histopathological report of the excised tumor confirmed the clinical and imaging diagnosis of cystic lymphangioma**. (Figure 13-14).** Patients recovered fully in post operative period without any complications. No

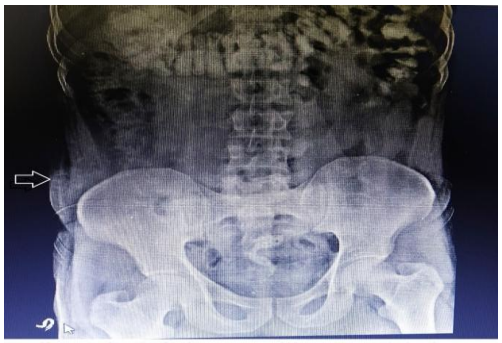
evidence of recurrence of the lesion was seen in the subsequent follow up visit up to eight weeks.



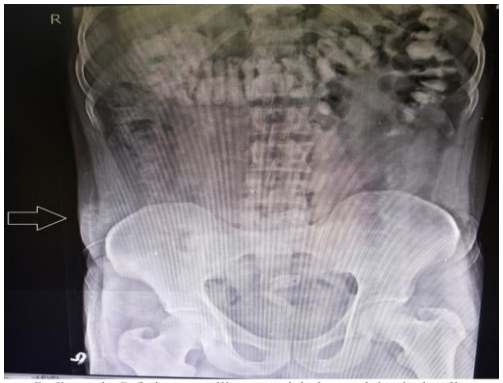
Swelling in right lower abdominal wall.
Figure 1



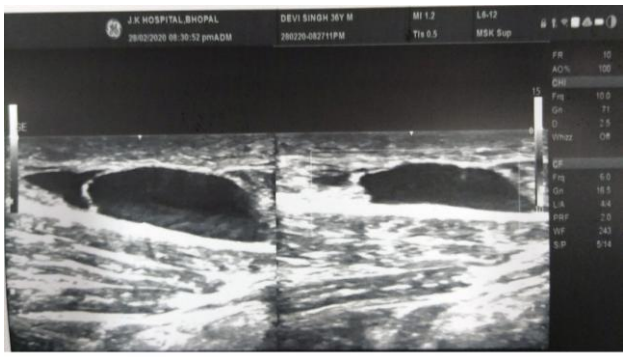
Swelling in right lower abdominal wall
Figure 2



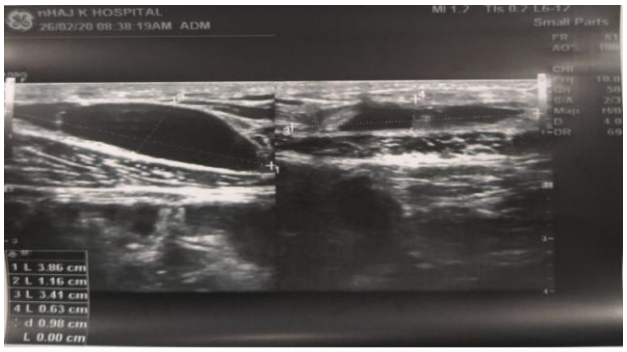
Radiograph- Soft tissue swelling over right lower abdominal wall
Figure 3



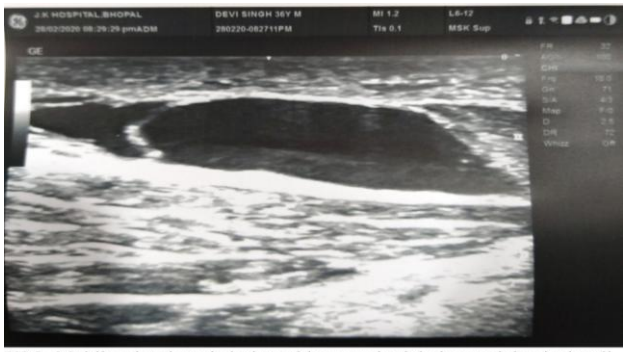
Radiograph- Soft tissue swelling over right lower abdominal wall.
Figure 4



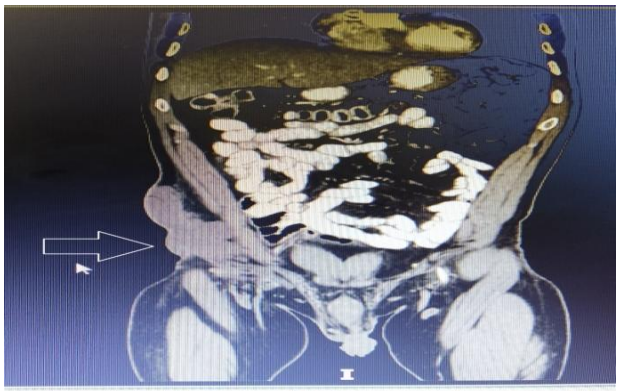
USG- Multiloculated cystic lesion with septae in right lower abdominal wall .
Figure 5



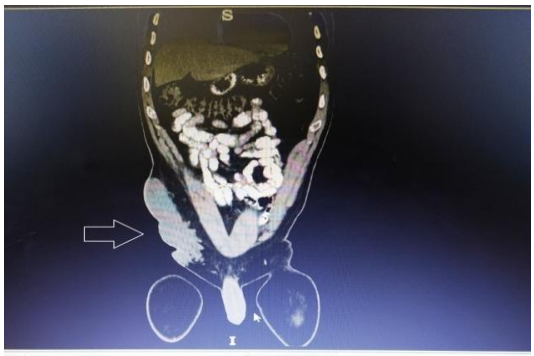
USG- Multiloculated cystic lesion with septae in right lower abdominal wall.
Figure 6



USG- Multiloculated cystic lesion with septae in right lower abdominal wall. No vascularity on colour doppler.
Figure 7

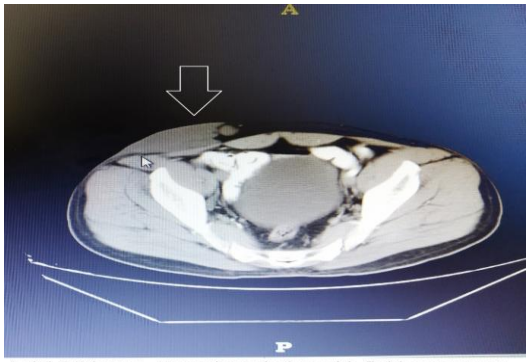


Coronal CT image- Hyperdense lesion with fluid component.(Arrow)
Figure 8



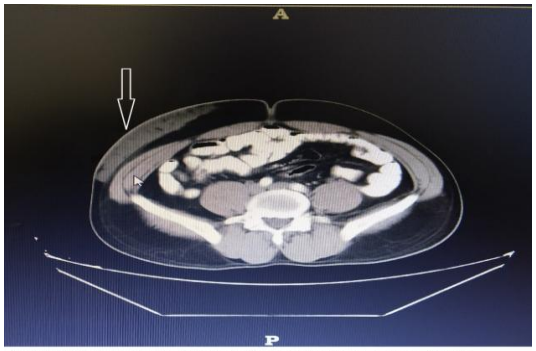
Coronal CT image- Hyperdense lesion with fluid component. (Arrow)

Figure 9



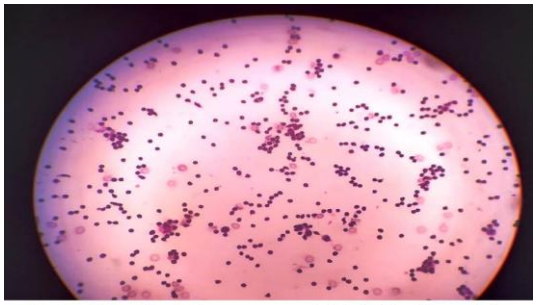
Axial CT image- Hyperdense lesion with fluid component. (arrow)

Figure 10



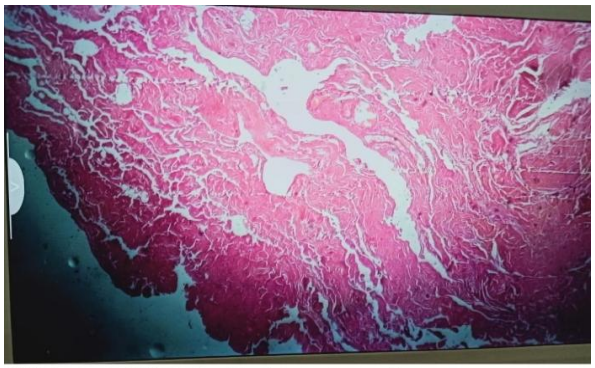
Axial CT image- Hyperdense lesion with fluid component.(arrow)

Figure 11



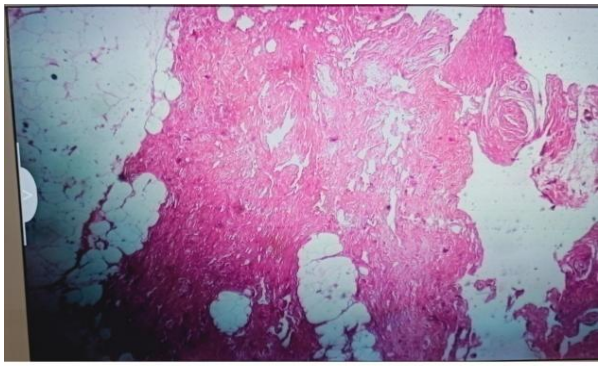
Cytological report S/O Cystic Lymphangioma.

Figure 12



HP image S/O Cystic Lymphnagioma.

Figure 13



HP image S/O Cystic Lymphangioma.

Figure 14

DISCUSSION

The lymphatic system develops as sac -like out growth from the endothelium of veins in the fifth week of gestation. There are six primary lymph sacs develops ,two in jugulars, two iliac, one retroperitoneal and one cisterna chyli. Failure of communication with lymph system results in formation of CL[6].

Cystic Lymphangioma or Hygroma is a congenital malformation of the lymphatic system and are of capillary, cavernous or cystic types. Also classified on basis of size of the cyst- as micro cystic(less than 2 cm), macro cystic (more than 2 cm in size) and mixed lymphangiomas (variable sizes of cysts) .Cystic lymphangioma / hygroma are results of sequestration of lymphatic tissue from lymphatic sacs during the development of lymphatic venous sacs. The sequestered tissue fail to communicate with the lymphatic or venous system and results in cystic dilatation of the lesion.[4-5]. Majority of cases present in the first 2 years of life and 65% are noted at birth. Wernher in 1843 described the various site of CL[6.]

Goldstein et al. focused on the prenatal diagnosis of CL during the 15th and 22nd week of gestation[2]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[3] . 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 site of occurrence are anogenital region, forehead, orbit, buttock, tongue, gastrointestinal tract, extremity,pelvis[6]. Hancock et al reported [3] various sites of involvement of CL are; 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%.Pandit et al [4] reported two cases of CL in the abdominal wall (4.4%).Very few cases of CL of abdominal wall were published so far[7-8] . In Kyeom Kim et al reported two case of CL, one was in the abdominal wall in LLQ and in other case CL was abdominal wall in RUQ[3].Ammar et al [7] reported a case of CL in 10 years female child in left side of abdomen with mass extending to pelvic cavity.

Riahinezhad et al [8] reported a case of 9 years male child with sudden onset of localized abdominal pain and bulging caused by rapid expansion of a hemorrhagic cystic lymphangioma of the abdominal wall. Most common presentation is slow growing painless lump in otherwise asymptomatic child. The effect of these lesions depends on their position and relationship to surrounding structure[4-5].In our case the lesion was slowly increasing in size, although parents noticed this in infancy. Ravi Kapoor et al described four different types of sonographic features for lymphangioma (a) cystic with thin septae;(b) cystic with thick septae;(c) cystic with thick septae and solid areas;(d)mainly solid with scattered cystic areas[9].

Anupam Lal et al described various radiological manifestations for abdominal lymphatic malformation[10]. Dave P K et al reported a case of C L of abdominal wall in a child [14]

Ultrasound study revealed a multiloculated cystic mass with thin internal septae and absent blood flow on color Doppler .CT finding are useful in determining the form, extent and nature of lesion [12]. USG and CT findings were similarly observed in our case. CT study reveals a large thin wall multiseptate cystic mass and show fluid attenuation, unless complicated by hemorrhage [11].However in our case MRI study was not performed.

Aspiration cytology of the lesion was showing predominantly lymphocytes against a proteinaceous background. 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 and infection, hypertrophied scar and lymphatic discharge from the wound[13]. About 20% of cases are reported to have recurrence of lesion .Post operative period was unremarkable and no recurrence of the lesion seen in follow up examination in our case[5].It is important to know the relationship between the cystic mass and the peritoneum as this decide the lesion is extra peritoneal in subcutaneous plane or with extension to peritoneal cavity. Therefore, it is important to differentiate between CL involving the preperitoneal fat layer and other cystic masses in peritoneal cavity. The presence of peritoneal lining elevation and prominent preperitoneal fat surrounding the mass can help in determining whether the mass is located within or in extra peritoneal cavity. These are valuable radiological observation and helpful for planning the surgery. In our case the lesion was extra peritoneal in subcutaneous planes of right lower abdominal wall. Use of sclerosing agents, laser therapy, radiation therapy and chemotherapy may be used as alternative treatment in poor surgical cases.

CONCLUSION

Cystic Lymphangioma / Hygroma is a commonly seen in pediatric age group, But unusual in adult. Abdominal wall is uncommon site of involvement , should be kept in differential diagnosis of cystic lesions. Various imaging findings are characteristic to diagnosed CL pre operatively. As standard protocol USG examination followed by CT/MRI examination used for proper evaluation in suspicious case of CL, Also useful for pre operative assessment of extension of lesion. Surgery is the mainstay of treatment if suitable.

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Conflicts of Interest -There are no conflicts of interest.

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