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INTRA-ABDOMINAL CYSTIC LYMPHANGIOMA IN A 30-YEAR-OLD MALE: A CASE REPORT AND REVIEW OF MANAGEMENT STRATEGIES

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ABSTRACT

Intra-abdominal cystic lymphangioma (ACL) is an uncommon cystic tumour of the abdomen and is considered a rare subtype of lymphangioma. These cystic tumours are mostly found in children, and the lesions are the result of an underdeveloped lymphatic system. Patients may have persistent abdominal pain and a noticeable abdominal mass. Non-invasive imaging, such as an ultrasound, as well as computed tomography scan can confirm the presence of a mass, but the final diagnosis is obtained through histopathological examination. Given the rare nature of ACL, there is limited information available, but the available literature suggests that complete surgical treatment is the best management. Searching aggressively will allow an adult patient to resolve the problem of this underreported disease in a more effective and timely manner.

KEYWORDS

Retroperitoneal cystic lymphangioma, abdominal pain, surgical excision

LEARNING POINTS

- Retroperitoneal cystic lymphangiomas are an exceptionally rare subset of benign lymphatic malformations in adults, often presenting with vague and nonspecific abdominal symptoms that complicate early recognition.
- This case of a 30-year-old male with retroperitoneal cystic lymphangioma underscores the diagnostic challenges, particularly when routine endoscopic and laboratory evaluations are unremarkable whereby contrast-enhanced computed tomography imaging proved pivotal in localizing and characterizing the lesion, informing surgical planning.
- This case highlights the necessity of maintaining a high index of suspicion for retroperitoneal cystic lymphangiomas in adults presenting with unexplained abdominal pain and reinforces the critical role of imaging and surgical management in achieving favourable outcomes.

INTRODUCTION

Cystic lymphangiomas (CLs) are rare, benign vascular malformations of the lymphatic system, histologically characterized by multiloculated cystic spaces containing lymphatic fluid. These lesions are believed to arise from congenital anomalies of lymphatic development, resulting in

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regional lymphatic obstruction during embryogenesis. While CLs predominantly occur in the cervical region and axilla, they have been reported in atypical locations, including the retroperitoneum, mediastinum, mesentery, omentum, colon, and pelvis, with perirenal involvement representing an exceptionally rare entity[1].

Intra-abdominal CLs in adults are exceedingly uncommon, with a reported incidence ranging from 1 in 20,000 to 1 in 250,000 individuals[1]. These lesions are frequently identified incidentally during imaging studies performed for unrelated conditions. Due to their rarity and nonspecific clinical presentation, they pose a significant diagnostic challenge and may mimic other intra-abdominal masses, such as mesenteric cysts or pancreatic neoplasms[1,2].

Recent case reports have documented incidental primary intra-abdominal CLs in adult patients, emphasizing the importance of including these lesions in the differential diagnosis of abdominal masses[1]. In addition, a case series of 32 adult patients with intra-abdominal CLs highlighted the diagnostic complexity and outlined principles for optimal management[3]. Rare presentations, such as giant pancreatic cystic lymphangiomas, further demonstrate the anatomical and clinical variability of these lesions[4].

Given the diagnostic challenges and potential for misidentification, a high index of clinical suspicion is warranted when evaluating intra-abdominal masses. Further studies and case reports are essential to enhance understanding of the epidemiology, clinical behaviour, and evidence-based management strategies for intra-abdominal CLs in adult populations [3].

CASE DESCRIPTION

A 30-year-old male with no significant past medical history presented with severe, non-specific abdominal pain. On physical examination, the abdomen was unremarkable with no palpable masses or organomegaly, but the patient reported severe pain. Initial workup included a gastroscopy, which was unremarkable, and a colonoscopy that revealed a benign 1 cm polyp not accounting for his symptoms. Due to persistent abdominal pain, a contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis was performed. The study revealed a 4.6 × 3.3 × 5 cm cystic lesion in the left retroperitoneal perinephric region with lobulated contours and fluid density (mean density 15 HU), without calcification or enhancing solid component, findings most consistent with a retroperitoneal lymphangioma *(Fig. 1)*. Additional findings included a nonobstructive 9 × 14 mm stone in the right lower calyx, a 7 mm simple cortical cyst in the left kidney, and a small fat-containing umbilical hernia. The liver showed mild diffuse fatty infiltration, while other abdominal organs including the spleen, pancreas, adrenal glands, bowel, and urinary bladder appeared unremarkable. There was no evidence of ascites, lymphadenopathy, or aggressive bone lesions. Overall, the imaging findings suggested that the retroperitoneal cystic lesion was the most likely aetiology behind the patient's abdominal pain.

DISCUSSION

Intra-abdominal cystic lymphangioma (ACL) is an unusual benign growth of the lymphatic system found within the abdominal cavity, representing fewer than 5% of all lymphangiomas, and is even rarer in adult populations; the majority of documented cases involve children, with lesions usually seated in the head, neck, or axilla. Consequently, an abdominal presentation in older individuals is an atypical and striking clinical observation[5,6]. Although the precise mechanisms underlying ACL are not fully elucidated, the literature presents two prevailing hypotheses: either a primary defect in lymphatic development or a later, acquired obstruction of lymphatic drainage. The congenital hypothesis argues that ACL arises from the persistence of embryologic lymphatic tissue that never opens to the central lymphatic circuit, causing isolated, cyst-like cavities to enlarge over time; the high frequency of diagnosis in children supports this model[6] The acquired obstruction model, however, accommodates adult cases better, positing that discreet inciting insults—including trauma, earlier abdominal operations, infectious processes, inflammatory conditions, and, on occasion, radiation—can provoke lymphatic occlusion or hyperplastic response, culminating in a cystic lesion. Supporting this view, Saxena et al.[7] reported that a considerable fraction of adults had a prior history of abdominal surgery, after which the lesion appeared. Regardless of the underlying cause, the lesion evolves into a benign, slowly expanding mass composed of enlarged lymphatic channels, each lined with a single layer of flattened endothelial cells, embedded within a framework of fibrous stroma mixed with scattered lymphoid aggregates. D2-40 and CD31 immunohistochemical staining reliably demonstrates the lymphatic origin of these lesions, thereby aiding differentiation from other cystic tumours within the abdomen[6]. On physical examination, abdominal lymphatic cysts present with a variable and often vague symptom profile, complicating early diagnosis. Abdominal pain as a single symptom dominates, being reported by up to 75%

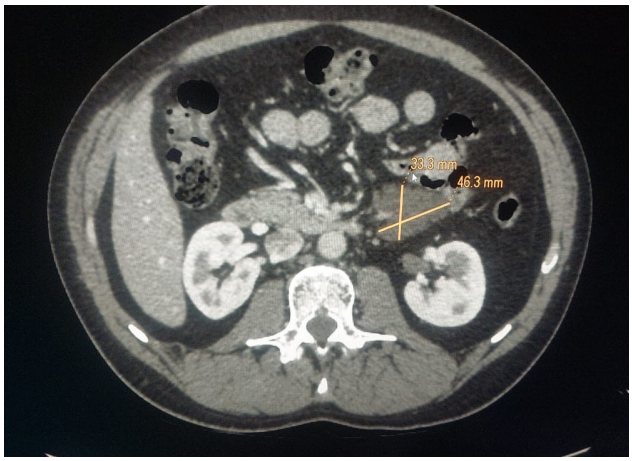


Figure 1. Contrast-enhanced axial computed tomography scan of the abdomen showing a well-defined retroperitoneal cystic lesion measuring approximately 46 × 33 mm, located adjacent to the left renal hilum.

of patients; this is accompanied by abdominal distention, a discernible mass, or rather diffuse gastrointestinal symptoms including nausea, vomiting, and altered bowel habits [3]. ACL may also be detected incidentally during imaging studies that are being conducted for entirely different complaints, emphasizing the potential for increasingly silent progression between scans. Although such cases are less frequent, ACL may be complicated by bowel obstruction, mesenteric volvulus, cystic haemorrhage, or superinfection, or, in the worst scenario, rupture. which can precipitate acute abdominal emergencies. Imaging tests remain central to establishing the diagnosis, starting with ultrasonography, which usually shows a cystic, anechoic lesion that may have single or multiple compartments. Further analysis with CT scans shows that these lesions are well-defined, thin-walled, hypodense cystic masses demonstrating fluid attenuation, occasionally revealed to have thin septations. After intravenous contrast injection, the cyst wall or septa may enhance, while calcifications are rarely present. Although magnetic resonance imaging plays a secondary role, its enhanced soft-tissue resolution helps in surgical planning by clearly mapping the cyst's relationship to adjacent structures and vessels[6]. The diagnosis is confirmed by histopathology after surgical excision; this step distinguishes the lesion from other cystic neoplasms, including mesenteric cysts, cystic mesothelioma, and some cystic tumours with malignant potential[7]. Treatment of adult lymphangioma depends on surgical removal, since watchful waiting is burdened by high rates of recurrence. Total excision or cystectomy is the preferred intervention because it consistently reduces the risk of recurrence and provides a definitive resolution of disease[5]. In a cohort of 32 adult patients in Tunisia 87% complete excision was achieved, and follow-up revealed a 9.4% recurrence rate, underscoring the critical need for radical resection. Analysing a 2012 dataset, the recurrence rate was anchored at 10% for cases where excision was deemed incomplete[5]. Surgical access options, namely laparotomy and laparoscopy, maintained comparable effectiveness; laparoscopic intervention is gradually preferred. The method is praised for diminished morbidity, quicker convalescence, and durable cyst-free periods, as corroborated by subsequent longitudinal findings. Indeed, a 2020 retrospective trial, predominantly laparoscopic, indicated sustained cyst clearance and satisfactory symptom control with negligible morbidity[5]. Alternatives such as aspiration, marsupialization, or circumferential partial resection, while occasionally entertained in complex scenarios threatening surrounding structures, yield recurrence rates exceeding 50% and appear to amplify operative morbidity[5]. Consequently, they should carry a presumptive "do not use" label whenever total excision remains executable. Furthermore, in a few cases clinicians have administered alcohol or acetic-acid guided sclerotherapy; yet pooled adult data reveal erratic effectiveness, and a lack of controlled trials relegates these modalities to investigational brackets, precluding their formal application in adult care. The prognosis following complete surgical excision of ACL remains highly favorable, as these lesions are benign and do not demonstrate malignant behavior; nonetheless, recurrence is not entirely ruled out, particularly in settings of partial resection or when comprehensive surveillance is neglected. Consequently, in patients with such lymphangiomas, particularly those who had incomplete excision, extended postoperative imaging follow-up is warranted[5].

ACL in the adult population is an uncommon yet harmless abnormality of the lymphatic system, presenting with ambiguous, uncharacteristic symptoms and displaying heterogeneous imaging characteristics on radiography. Definitive diagnosis hinges upon histological examination. The disease mechanism may either be congenital or acquired, while surgical resection remains the benchmark of treatment. When resection is performed entirely via laparotomy or laparoscopy, excellent outcomes with a particularly low recurrence rate are achievable; in contrast, incomplete excision markedly amplifies the probability of subsequent relapse[5]. Procedures that do not involve surgical resection have been the object of investigation, yet their established utility remains negligible, with amassed data compellingly favouring surgical excision as the primary form of management. Consequently, when faced with a case of a cystic abdominal lymphangioma presenting in adulthood, the rarity of the diagnosis poses a distinctive clinical difficulty. Prompt identification of characteristic clinical, imaging, and histological features, alongside application of the surgical standards outlined, translate into strongly favourable prognoses for those affected[3,5,6].

CONCLUSION

Retroperitoneal cystic lymphangiomas represent an exceptionally rare subset of benign lymphatic malformations in adults, often presenting with vague and nonspecific abdominal symptoms that complicate early recognition. Our case of a 30-year-old male underscores the diagnostic challenges, particularly when routine endoscopic and laboratory evaluations are unremarkable. Contrast-enhanced CT imaging proved pivotal in localizing and characterizing the lesion, informing surgical planning. Definitive diagnosis requires histopathological confirmation, and complete surgical excision remains the treatment of choice, providing durable symptom relief and minimizing recurrence risk. This case highlights the necessity of maintaining a high index of suspicion for retroperitoneal cystic lymphangiomas in adults presenting with unexplained abdominal pain and reinforces the critical role of imaging and surgical management in achieving favourable outcomes.

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