Chylous ascites, genital and lower-limb lymphoedema: a case report

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Key words

Chylous ascites, Decongestive lymphatic therapy, Genital lymphoedema, Lower-limb lymphoedema, Thoracic duct stenosis, Total parenteral nutrition

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Abstract

There is evidence in the literature of clinicians dealing with either lymphoedema or chylous ascites (CA), but associations between these conditions are rarely discussed. This paper aims to highlight the importance of teamwork and information exchange in the multidisciplinary approach of a case of CA with lymphoedema. A seventy-four-year-old woman was referred to the Lymphoedema Unit with bilateral lower-limb lymphoedema (LLL) and genital lymphoedema. She also presented with signs of protein malnutrition secondary to abdominal CA. The prescribed mainstream treatment was total parenteral nutrition (TPN), decongestive lymphatic therapy with manual lymphatic drainage and multi-layer short-stretch bandages of the lower limb. This management provided a satisfactory outcome. The association of CA and lymphoedema often presents as a complex clinical case, which requires a multimodal treatment based on proper nutrition regimens and decongestive therapy. Effective teams are crucial to achieving best practice in such challenging scenarios.

Chylous ascites (CA) is an uncommon finding, defined as the leakage of the lipid-rich lymph into the peritoneal cavity. It often represents as a consequence of the therapeutic strategies for solid tumour management, which includes radiotherapy and surgery. Even though the reported incidence of CA is 1/20,000 admissions overall, the increased use of such aggressive strategies and the increased survival rates of patients with cancer may be making such occurrences more common (Al-Busafi et al, 2014).

Lymphoedema can be associated with CA, since lymphatic drainage is usually compromised. This entity is defined as an abnormal accumulation of interstitial fluid, due to a decreased lymphatic uptake, with subsequent local and systemic inflammatory response that eventually leads to swollen tissues. In particular, genital lymphoedema and lower-limb lymphoedema (LLL) are common findings correlated with CA (Brouillard et al, 2014).

The authors present the case of a patient with rectal cancer who developed severe CA and LLL secondary to an idiopathic thoracic duct stenosis, which worsened after radiotherapy. This case study aims to highlight the importance of a correct diagnosis of this disease and of how a multidisciplinary approach can help to achieve good outcomes.

Case report

The patient was a 74-year-old female referred to the Lymphoedema Unit with bilateral LLL and genital lymphoedema of 30 years' duration.

She previously underwent rectal resection with direct colorectal anastomosis for G2 rectal adenocarcinoma (pT3, pN0, M0) and received adjuvant chemotherapy and radiotherapy. CA was an occasional finding during surgery and remained untreated. Two years ago, a 10-litre paracentesis was performed for tension ascites. The liquid cytology was negative for tumoral cells and triglyceride levels were 1,453 mg/dL. Subsequently, the course was uneventful except for persistent lymphoedema and residual mild CA, which did not require any specific treatment. At the time of the evaluation at the Lymphoedema Unit, the last oncological follow-up revealed no recurrence of tumoral disease. At physical examination, the patient presented with bilateral LLL, more prominent in the left limb, affecting the whole leg, from toes to the groin, with 6 mm of pitting sign. Stemmer's sign was positive bilaterally, the difference of perimeter on the calves was 7 cm by tape measurements. In addition,

dermal thickening and dyschromia of the skin were observed. Vulvar lymphoedema was also detected. She was diagnosed with Stage 3 chronic genital lymphoedema as she had redundant, orange peel skin in the pubis and labia majora.

The patient's abdomen was distended and there was dullness to percussion over all the abdomen, except for the epigastrium and in the right upper quadrant. The patient presented shifting dullness, compatible with ascites.

Figure 1 shows the main findings of the physical examination. Blood tests including nutritional assessment and lymphoscintigraphy were ordered. Blood test results showed significant hypoproteinemia, with decreased levels of serum albumin and total proteins (*Table 1*). Lymphoscintigraphy (*Figure 2*) revealed no sign of migration, 10 minutes after introducing the tracer. Three-and-a-half hours later, a second image showed minimum uptake in the derma over the distal limbs and the right groin. Twenty-four hours later, an abdominal image was obtained and no accumulation of activity was detected due to tracer leakage. The authors concluded along with the radiology team that the patient probably had a primary lymphoedema caused by an idiopathic thoracic duct stenosis, worsened by radiotherapy.

To manage both problems, a multidisciplinary approach was needed. In fact, if we had treated only LLL with the decongestive lymphatic therapy (DLT), the CA could have worsened, as lymphatic fluid would have accumulated in the abdomen due to the dysfunction of the lymphatic system.

The cause of CA was still uncertain, as the site of the leakage had not been determined by the lymphoscintigraphy team. Even though an oral contrast test (a fat-containing test meal) and subsequent laparoscopy could have detected the chylous leakage, and possibly allowed to close it surgically, a conservative approach was attempted first.

In order to treat the severe protein-calorie malnutrition, the patient was admitted to the Endocrinology and Nutrition Department. She received TPN for 12 days. The initial TPN had 12 g of proteins and 1,530 kilocalories, that was later tailored to 16 g of proteins. During the hospital stay, DLT was performed at the same time, with manual lymphatic drainage and multi-layer short-stretch bandages of the lower limb to reduce the oedema of the limbs.

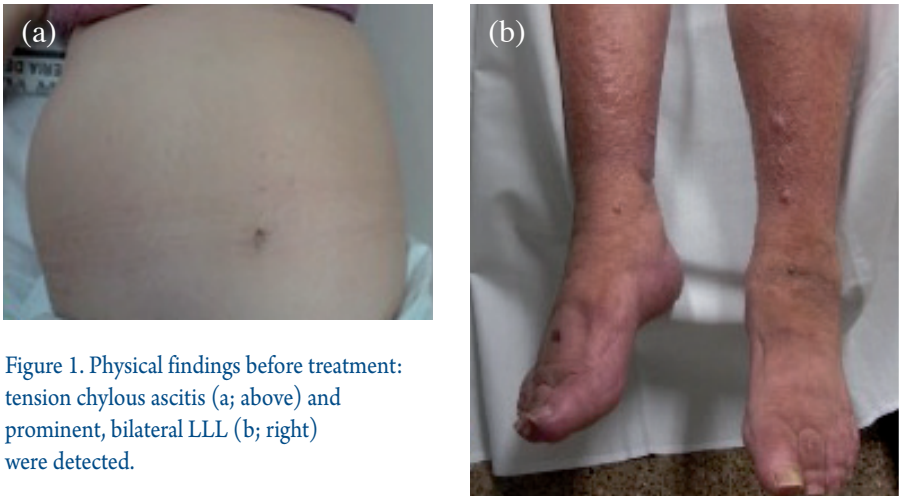

Figure 1. Physical findings before treatment: tension chylous ascitis (a; above) and prominent, bilateral LLL (b; right) were detected.

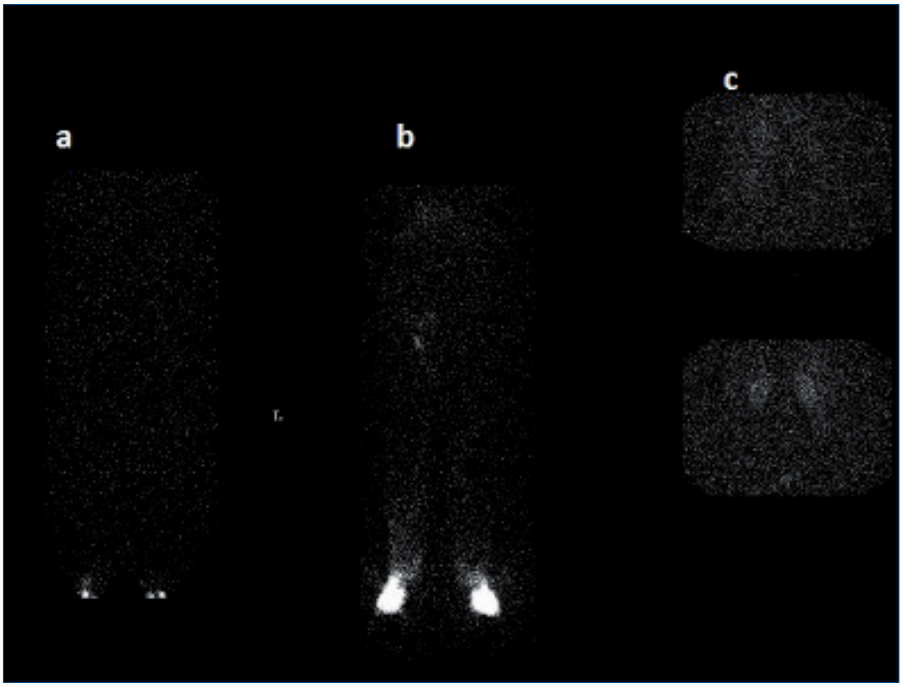

Figure 2. Lymphoscintigraphy: a) first image showed no sign of migration ten minutes after introducing the tracer b) three and a half hours later, a second image showed minimum uptake in the derma over the distal limbs and the right groin c) twenty-four hours later, abdominal images were obtained and no accumulation of activity was detected.

The progress was satisfactory, resulting in resolution of CA and an improvement in LLL. During her hospital stay, the patient lost 9 kg — this was probably due to multifactorial causes, such as muscle loss due to inactivity during her stay at the hospital, an episode of diarrhea and absorption of lymphatic fluid due to successful treatment. There was also a significant improvement in the nutritional parameters. *Table 1* summarises the main clinical findings at admission and at discharge.

To maintain the results of the treatment after discharge, a multidisciplinary approach was planned. The Nutritional Department prescribed a low-fat maintenance diet, four spoonfuls of middle-chain triglyceride oil (MCT) a day, protein modules, K+, Ca++ and vitamin D supplements to avoid the relapse of CA. A flat-knitted class 3 garment was prescribed by the Lymphoedema Unit to preserve the reduction of lymphoedema.

Two months later, the patient was reevaluated. She had followed the

Table 1. Clinical findings at admission and at discharge.

	At admission	At discharge
Chylous ascitis	Present	Absent
Weight	65 kg	52 kg
Albumin	1.97 g/dL	2.9 mg/dL
Total proteins	4.6 g/dL	6.4 g/dL

treatment prescribed at discharge, and both CA and LLL were correctly controlled.

Discussion

Chylous ascites is a rare clinical condition that occurs as a result of disruption of the abdominal lymphatics. When associated with lymphoedema, as in this case, the management of both of the diseases can be challenging.

Chylous loss carries several clinical consequences, such as the decrease of essential proteins, lipids, immunoglobulins, vitamins, electrolytes and water. Furthermore, it can lead to hypogammaglobulinemia, increasing the risk of infections (Al-Busafi et al, 2014). Abdominal distension, pain, diarrhea, dysphagia and peripheral oedema are the most frequent associated symptoms and often considered a direct consequence of CA (Lizaola et al, 2017).

Lymphoedema should be considered a part of the clinical picture, which can be associated with CA, and a multidisciplinary approach must be applied in its management when detected. Knowing its aetiology is important for the correct management. Lymphatic disease can be primary or secondary to infection, surgery or radiotherapy (Brouillard et al, 2014; Zasadzka et al, 2018). Similarly, CA can be traumatic (due to surgery or radiotherapy) and atraumatic (congenital, inflammatory or systemic disorders; Lizaola et al, 2017).

Lizaola et al (2017) described the diagnosis of CA as clinical and supported by the biochemical findings of paracentesis. However, lymphoscintigraphy can be useful in detecting abnormal lymphatic drainage in CA, such as leakages and stenosis of the thoracic duct (Lizaola et al, 2017). In this case, all these diagnostic tools were used, obtaining the final etiology by lymphoscintigraphy. The therapeutic process of CA is complex and aims to correct the cause, to improve the comfort of the patient and to reduce the risk of recurrence.

A milestone of treatment consists of ensuring a correct nutritional status with high protein and low-fat diet, along with MCT supplement (Weniger et al, 2016; Lizaola et al, 2017). Lv et al (2017) reported on TPN being among the key therapeutic tools for CA as being particularly useful during the acute phase. It contributes lowering the lymphatic production by providing bowel rest, and helps to increase peritoneal effusion and plasma absorption by enhancing the oncotic pressure. Finally, Al-Busafi et al (2014) proposed abdominal paracentesis as an additional therapeutic option, as it mitigates symptoms and can be repeated when needed. However, the risk of electrolyte imbalance, infections and malnutrition are possible complications (Lizaola et al, 2017). Another potential flaw is to create a negative pressure that may worsen the lymphatic leakage, causing the relapse of CA.

As for LLL, DLT is the treatment of choice and is effective in reducing the volume of lymphoedema. DLT is a combination of physical therapies, manual lymphatic drainage and multilayer bandages to help draining the lymph in the interstitial tissue. For long-term management of LLL a low-stretch garment must be worn daily (Zasadzka et al, 2018)

In the reported case, CA was successfully treated with the simple combination of a low-fat diet with MCT supplement and TPN. No somatostatin was administered, and only one abdominal paracentesis in tension ascites was needed. LLL improved satisfaction with inpatient DLT and subsequent garment maintenance. Interestingly, the combination of the specific treatments of the two entities led to a synergetic improvement of both conditions. At a root cause analysis, the presence of an underlying primary lymphatic dysfunction with chronic LLL made this case particularly challenging. A proper diagnosis of the thoracic duct stenosis, if known before radiotherapy, might potentially have prevented CA.

Conclusion

The association of CA and lymphoedema constitute a complex clinical presentation that will benefit from a multidisciplinary treatment, based on a nutrition regimen associated with decongestive therapy of LLL. A multidisciplinary team approach is crucial for a successful outcome in this unique situation.

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