



Intestinal Lymphangiectasia: Insights on Management and Literature Review

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Statistical Analysis C
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Manuscript Preparation E
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Patient: Male, 24
Final Diagnosis: Intestinal lymphangiectasia
Symptoms: Frequent episodes of diarrhea • recurrent infections • swelling in the lower limbs
Medication: Octreotide • MCT oils
Clinical Procedure: Endoscopic exam • Doppler ultrasound study • abdominal CT scan
Specialty: Gastroenterology and Hepatology


Objective: Rare disease
Background: Intestinal lymphangiectasia (IL) is a rare disease characterized by a dilatation of the intestinal lymphatics and loss of lymph fluid into the gastrointestinal tract leading to hypoproteinemia, edema, lymphocytopenia, hypogammaglobinemia, and immunological abnormalities. Iron, calcium, and other serum components (e.g., lipids, fat soluble vitamins) may also be depleted. A literature search revealed more than 200 reported cases of IL. Herein, we report our observations of a patient diagnosed with IL; we also present our conclusion for our review of the published literature.

Case Report: A 24-year-old male was admitted to Aleppo University Hospital with the complaints of abdominal pain, headache, arthralgia, fever, and rigors. His past medical history was remarkable for frequent episodes of diarrhea, recurrent infections, and swelling in the lower limbs. In addition, he had been hospitalized several times in non-academic hospitals due to edema in his legs, cellulitis, and recurrent infections. In the emergency department, a physical examination revealed a patient in distress. He was weak, dehydrated, pale, and had a high-grade fever. His lower extremities were edematous, swollen, and extremely tender to touch. The overlying skin was erythematous and warm. Moreover, the patient was tachycardic, tacypneic, and moderately hypotensive. The patient was resuscitated with IV fluids, and Tylenol was administered to bring the temperature down. Blood tests showed anemia and high levels of inflammatory markers. The patient's white blood cell count was elevated with an obvious left shift. However, subsequent investigations showed that the patient had IL. Suitable diet modification plans were applied as a long-term management plan.

Conclusions: IL is a rare disease of challenging nature due to its systematic effects and lack of comprehensive studies that can evaluate the effectiveness of specific treatments in a large cohort of patients. Medium-chain triglyceride (MCT) oils and diet modification strategies are effective in reducing the loss of body proteins and in maintaining near-normal blood levels of immunoglobulins. However, octreotide and MCT oils had no proven role in reducing lymphedema in our patient.

MeSH Keywords: Lymphangiectasis, Intestinal • Octreotide • Protein-Losing Enteropathies • Cellulitis

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/899636>

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Background

Intestinal lymphangiectasia (IL) is a rare disorder characterized by impaired intestinal lymphatic drainage due to primary ectasia, diffuse or localized, of the intestinal lymphatic vessels, or as a result of secondary impacts on the normal lymph circulation, such as heart diseases and retroperitoneal lymph node enlargements [1]. Regardless of the cause, the pathogenesis of IL involves a dilation of the intestinal lymphatic vessels and an increase in the interstitial pressure, leading to lymph fluid leakage into the intestinal lumen. This outflow of lymph fluid decreases the plasma oncotic pressure due to a lack of proteins and gamma-globulins in the plasma. In addition, fat, lymphocytes, electrolytes, and fat soluble vitamins can be depleted [2,3].

The age of disease onset is variable. Although the diagnosis was established, in multiple studies, in infants. But IL has been observed in older age groups. It has been reported that the mean age of disease onset is twelve years. No etiological causes for IL have been identified, and genetic etiology is not clearly defined. Almost all the reported cases are of sporadic origin and familial stories of IL in first- or second-degree relatives are rarely reported [4]. Prevalence is unknown and disease distribution around the world is poorly researched due to the rareness of the disease.

The symptoms of IL are variable depending on its severity. The classical symptoms are bilateral or unilateral lower limb edema and intermittent diarrhea. Some patients develop steatorrhea; in other cases, an accumulation of fluids in body cavities resulting in pleural effusion or ascites has been reported [5,6]. The main patient complaints are abdominal pain, nausea and vomiting, intermittent diarrhea and steatorrhea, chronic fatigue, weakness, weight loss, and inability to gain weight.

IL is a challenging disease due to its physical characteristics and its psychological effects on the patients and their families. Diet modification plans, medium-chain triglycerides (MCT), octreotide, antiplasmin, and surgical procedures have been introduced as management choices for the disease [7–10].

Here, we present a case of a patient with IL. This patient was followed, closely, for more than three years. In addition, we searched the literature looking for similar cases of IL. The purpose of this report is two-folds: to present our case study and IL management plan and present the conclusions from an investigation of the literature.

Case Report

A 24-year-old male was admitted to Aleppo university Hospital (AUH), in 2010, with the complaints of abdominal pain, headache, arthralgia, fever, and rigors. The patient stated that he had,

since infancy, intermittent episodes of foul-smelling diarrhea. These episodes were accompanied by abdominal pain, nausea, and vomiting. The patient was suffering from intractable bilateral swelling in the lower limbs, more prominent in his left leg. This swelling was responsible for restricting his daily life activities.

The current hospitalization was one in a series of admissions to a non-academic hospitals, inside and outside the country. These admissions were for multiple reasons. Reviewing the patient's medical history revealed that recurrent infections were the most common cause for all of his previous hospitalizations. In the emergency department, a physical examination revealed a distressed patient with an ill appearance. He was weak, dehydrated, pale, and had a high-grade fever of 39.6°C (103.2°F). The patient was lying supine on the exam table with his legs drawn down, arms on his abdomen, and he was shivering and staring with an anxious facial expressions and eyes that were filled with tears. Moreover, the patient was dyspneic with a respiratory rate of 17 beats/minute and without cyanotic hue; his oxygen saturation was 92%. The patient was tachycardic (pulse rate, 125 beats/minute), and moderately hypotensive (blood pressure 97/60 mm Hg).

The patient's lower extremities were severely edematous, and extremely tender to touch. The overlying skin was erythematous and warm. The redness of the skin extended from the left popliteal area to the scrotum, which was enlarged but not tender to palpation. The skin covering the scrotum had changed to a very thick and hard skin mass. Further investigation revealed that the patient had an insect bite on the top of his left foot, one week ago.

The patient was resuscitated with IV fluids. Tylenol was infused to bring his temperature down, and to control pain. Blood tests in the emergency department showed a high C-reactive protein (CRP) level and an elevation in the erythrocyte sedimentation rate (ESR). White blood cell count was 18,500 per mm³ with 85% neutrophilia. Hemoglobin was 10.3 g/dL and the hematocrit was 31%. The patient was admitted to the general internal medicine department in AUH for further investigations and management.

In the general internal medicine department, the management plan was applied immediately, including IV fluids, empiric antibiotic therapy, and analgesics. Additional blood samples were drawn for further laboratory testing, blood culture, and antibiotic sensitivity tests. Table 1 presents a summary of the laboratory test results.

Doppler ultrasound study of the lower limbs and scrotum was performed and showed a diffuse thickening, increasing the echogenicity of the skin and subcutaneous tissues. These findings were accompanied by hypochoic strands of fluids that extended between the hyperechoic fat and the underlying

Table 1. Results of blood tests.

	Blood test	Results	Normal range
CBCs	WBC and differential	18,500 (N 85%–L 5%)	5–10×1,000/mm ³
	HGB	10.3	12–18 g/dL
	HCT	31	37–52%
	PLT	322	150–450×10 ⁹ /L
Inflammatory markers	ESR	43/72	mm/L
	CRP	32	mg/L
RFTs	Urea	45	7–20 mg/dL
	Creatinine	0.7	0.6–1.2 mg/dL
Electrolytes	Sodium	135	135–145 mmol/L
	Potassium	3.8	3.5–5.5 mmol/L
	Calcium	7.9	8.5–10.2 mg/dL
	Magnesium	1.7	1.5–2.5 mEq/L
LFTs	ALT/AST	45/35	7–56 unit/L
	PT	13.8	Seconds
	INR	1.1	–
Chemistry	ALP	110	44–147 IU/L
	Bilirubin (total/direct)	1/0.8	0.3–1.9 mg/dL
	Total protein	3.2	6–8 g/dl
	Albumin	2.1	3.5–5.5 g/dL
Immunology	ANA	Negative	<1/0 IU
	Anti-dsDNA	Negative	<1/0 IU
IG	IGG	35	650–1,500 mg/dL
	IGA	10	80–350 mg/dL
	IGM	14	45–250 mg/dL
	IGE	2	150–1,000 UI/mL
Thyroid function test	TSH	1.6	0.3–5.0 U/mL
	FT4	1.1	0.8–2.8 ng/dL
Lipid profile	Cholesterol	79	180–200 mg/dL
	LDL	80	100–129 mg/dL
	HDL	42	35–65 mg/dL
	TG	35	150–200 mg/dl

CBC – cell blood count; WBC – white blood cell; HGB – hemoglobin; HCT – hematocrit; PLT – platelet; ESR – erythrocyte sedimentation rate; CRP – C-reactive protein; RFT – renal function test; LFT – liver function test; ALT – alanine aminotransferase; AST – aspartate aminotransferase; ALP – alkaline phosphate; PT – prothrombin time; INR – international normalized ratio; ANA – antinuclear antibody; Anti-dsDNA – anti-double stranded DNA; IG – immunoglobulin; LDL – low-density lipoprotein; HDL – high-density lipoprotein; TG – triglycerides.

connective tissues. A characteristic “cobblestone” appearance was observed along the left leg. The veins in the left leg were non-compressible and Doppler flow was clearly sufficient. The vessels in the scrotum area were intact and the blood supply to the testes was adequate. An accumulation of scrotal fluids was observed bilaterally, extending slightly to the penis.

Abdominal sonogram revealed an edematous gallbladder wall and large bowel loops with markedly thickened walls. A slight amount of ascetic fluid was observed in the peritoneal cavity.

Three days later, when the patient clinically stabilized, further investigational procedures were planned to study the swelling of the lower limbs. The CT scan was very useful in identifying dilated intestinal loops and a diffuse thickening of the intestinal walls, as well as mesenteric edema. The CT scan showed that the circumferential thickening of the small bowel wall was enlarged (1.5–2 cm).

One week later, an endoscopic examination was performed, which was significant for mucosal changes and white spots and nodules (dilated lacteals) (Figure 1A). Multiple biopsies were obtained for further histological examinations, which showed a characteristic dilatation of the enteric lymph vessels in the mucosal and submucosal layers without any evidence of inflammation. The histological examination was suggestive of IL (Figure 1B).

The results of our investigations, integrated with the patient’s clinical picture, were suggestive of an IL associated with cellulites in the left lower limb.

Three weeks later, the patient was totally recovered; he was discharged home and scheduled for a routine follow-up visits in the internal medicine clinic at AUH.

The patient was followed for more than three years. His long-term management plan was designed to primarily educate the patient and his parents about the specifics of his disease. In addition, the management included dietary modifications, routine screening tests, and some medications to control his chronic diarrhea. The patient was advised to use a compression stocking, raise his leg while in a supine position, and to do routine massages to the lower limbs.

While the patient was under observation in the hospital, octreotide was applied subcutaneously, and we continued this treatment with a slow-release formulation for a period of six months.

The most important part of our management plan was diet modifications. The nutritionist in our clinic had educated the patient to substitute the long-chain fatty acids (LCFA) in his food with medium-chain triglycerides (MCTs). Due to its availability and cheapness, the patient started to consume palm oil as a per-oral morning load and in his daily meals. He followed the palm oil diet for seventeen months. The palm oil diet was replaced later with a coconut oil diet. See (Figure 2) for clinical features of IL in our patient.

In order to track the effectiveness of the previously mentioned procedures on the amount of swelling in the lower limb, we measured the diameter of his leg and the middle part of his thigh. The anatomical landmarks for these measures were

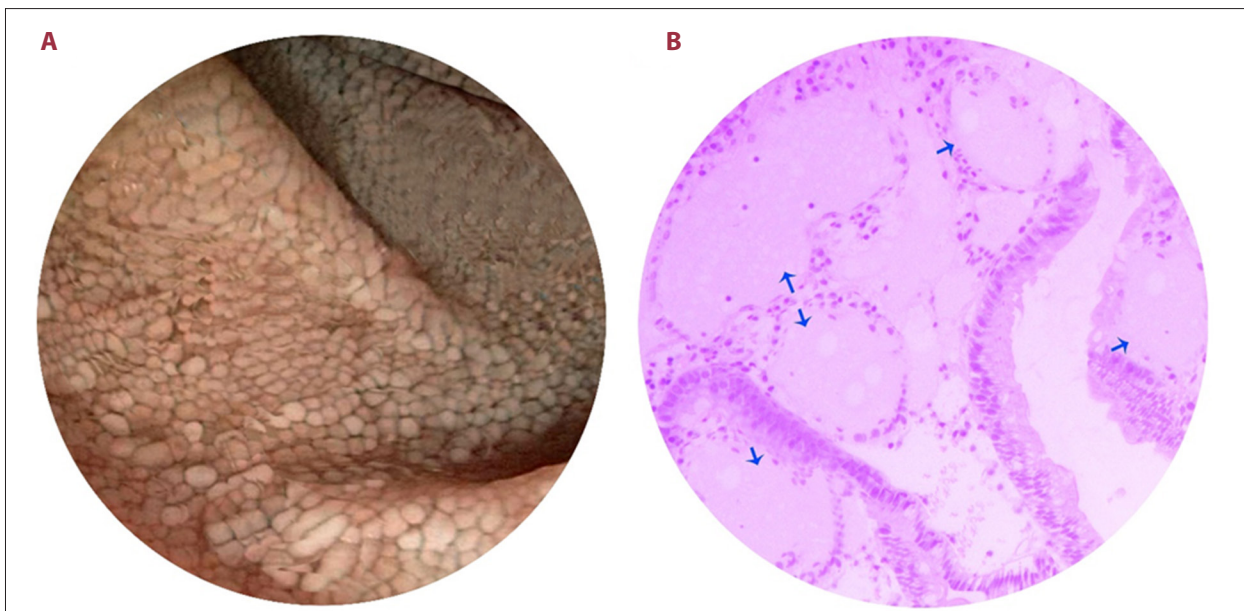


Figure 1. Endoscopic and histological findings. Mucosal changes showing white spots and nodules (dilated lacteals) in the duodenum (A) and dilated lymphatic vessels (blue arrows) (B).

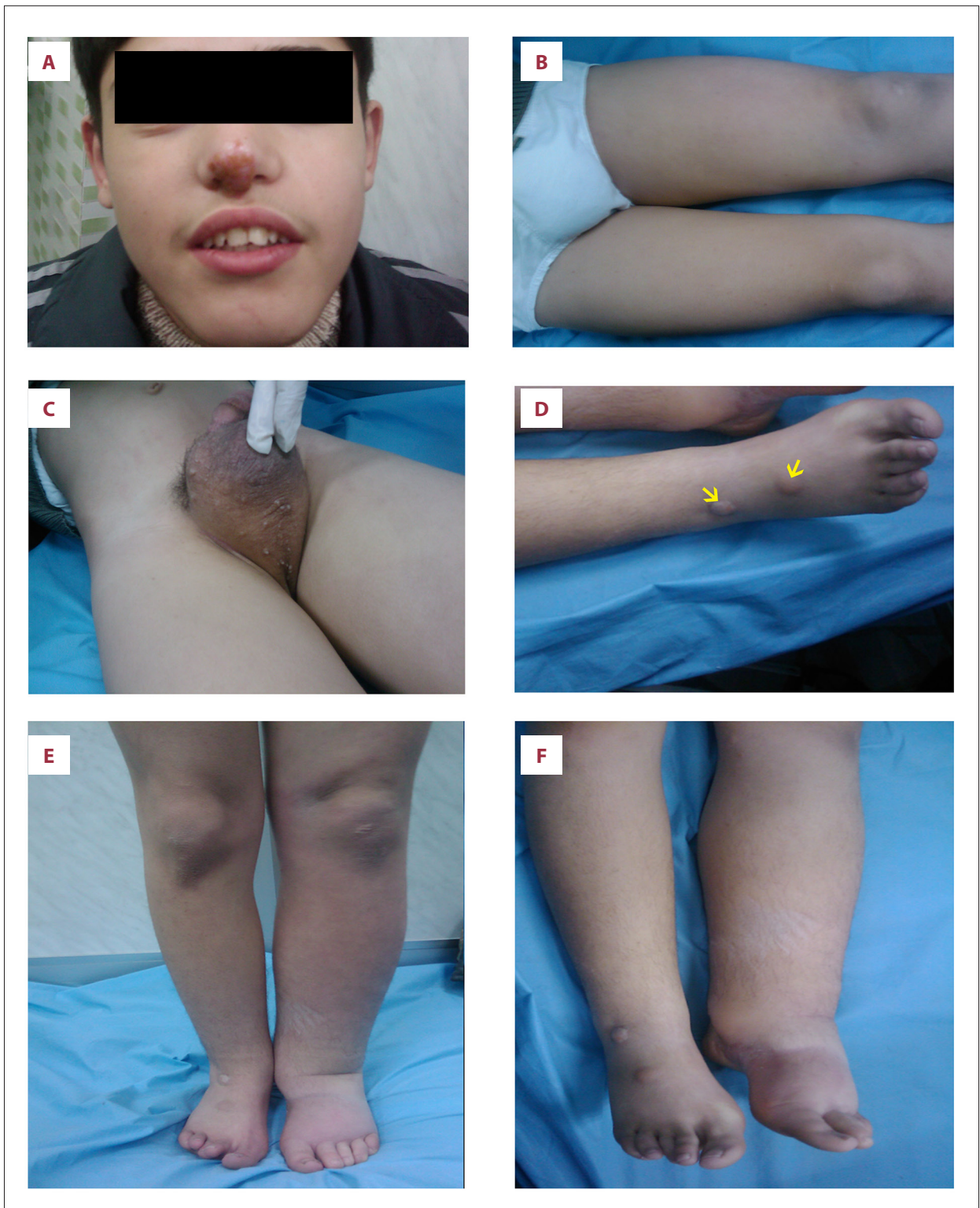


Figure 2. Clinical findings of intestinal lymphangiectasia. The patient showing yellow scab on the top of the nose due to infection (A); Lower limb edema extends to the thigh (B); Scrotal skin thickening (C); Warts (yellow arrows) on the dorsal part of the right foot (D); Edema after discharge from the hospital (E); Edema after two years of management (F).

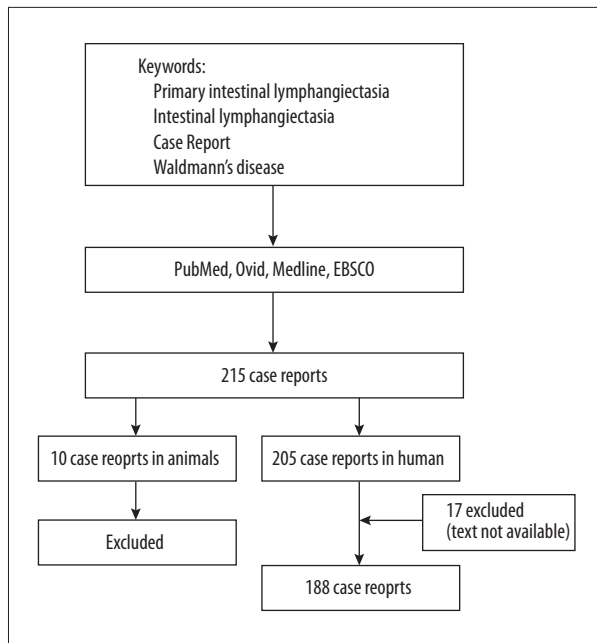


Figure 3. Flowchart for literature search showing the included and excluded articles.

the middle part of the thigh, the lateral malleolus of the fibula, and the medial malleolus of the tibia. We recorded these measurements during every visit and at the same time of day (i.e., the patient was scheduled at 13:00 PM and his measurements were taken at 13:30 PM).

Practically speaking, we have never seen any significant improvement in the size of the swelling in the lower limb or in the scrotal area. It seems that the regression of edema in the lower leg was slightly better when coconut oil was applied, but this change was trivial overtime. Notably, the edema was less in the early morning and greater in the evening. The mechanical maneuvers (e.g., raising the leg during sleep, massages) were temporarily effective in reducing the edema size.

The diet modification plan was of great benefit in preventing further accumulation of fluids, controlling the loss of enteric proteins, and in preventing susceptibility to infectious agents. The patient was never rehospitalized for any reason up to the time of drafting this article. MCT oils had no important effects on edema size in the long-term management plan for our patient.

Discussion

We performed our literature search in PubMed using the keywords “primary intestinal lymphangiectasia, intestinal lymphangiectasia, Waldmann’s disease, case report”. Feeding the same words into Ovid, Medline, and EBSCO led to the same results. Two hundred and fifteen articles were identified. Two

of the co-authors reviewed the literature separately, and they matched the results. According to our search, the first case report of IL was published in 1965 [11]. Ten articles described IL in animals (nine articles were for a diagnosis of IL in dogs and one in a horse. One paper was a pharmaceutical experiment on a guinea pig [12]). The remaining 205 articles described IL in humans.

The original text was not available for seventeen articles, so they were excluded from our review. The final number of articles included in our review was 188 studies in humans. Figure 3 presents a flowchart for our literature search.

The articles were classified as follow:

1. Articles described a single case report and its clinical course in a patient.
2. Articles presented multiple case reports and its clinical manifestations in each patient.
3. Articles studied an association between IL and other etiologies.
4. Articles dedicated their attention to the complications of IL.
5. The role of specific treatments in the management of IL was presented in multiple studies; these treatments were: octreotide, MCT oil, low fat diet plans, and anti-plasmin.
6. Some studies were interested in an investigational means to establish the diagnosis of IL.

Literature search revealed that the age at diagnosis was widely variable. While some cases were diagnosed early in life, others were detected later in life. Age was clearly elucidated in 180/188 case reports. Figure 4 present the number of case reports in each age group. The total number of male and female patients included in the reported cases (178/188) was 100 males and 78 females. Sporadic IL cases were more prominent; almost 90% of the reported IL cases were of sporadic origin and the rest were of familial susceptibility with a genetic basis.

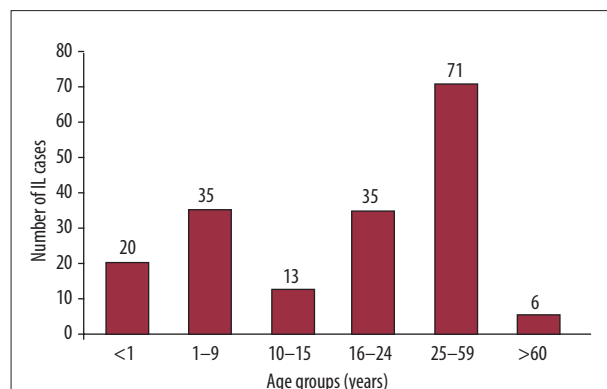


Figure 4. Column chart showing the number of reported case reports within each age group.

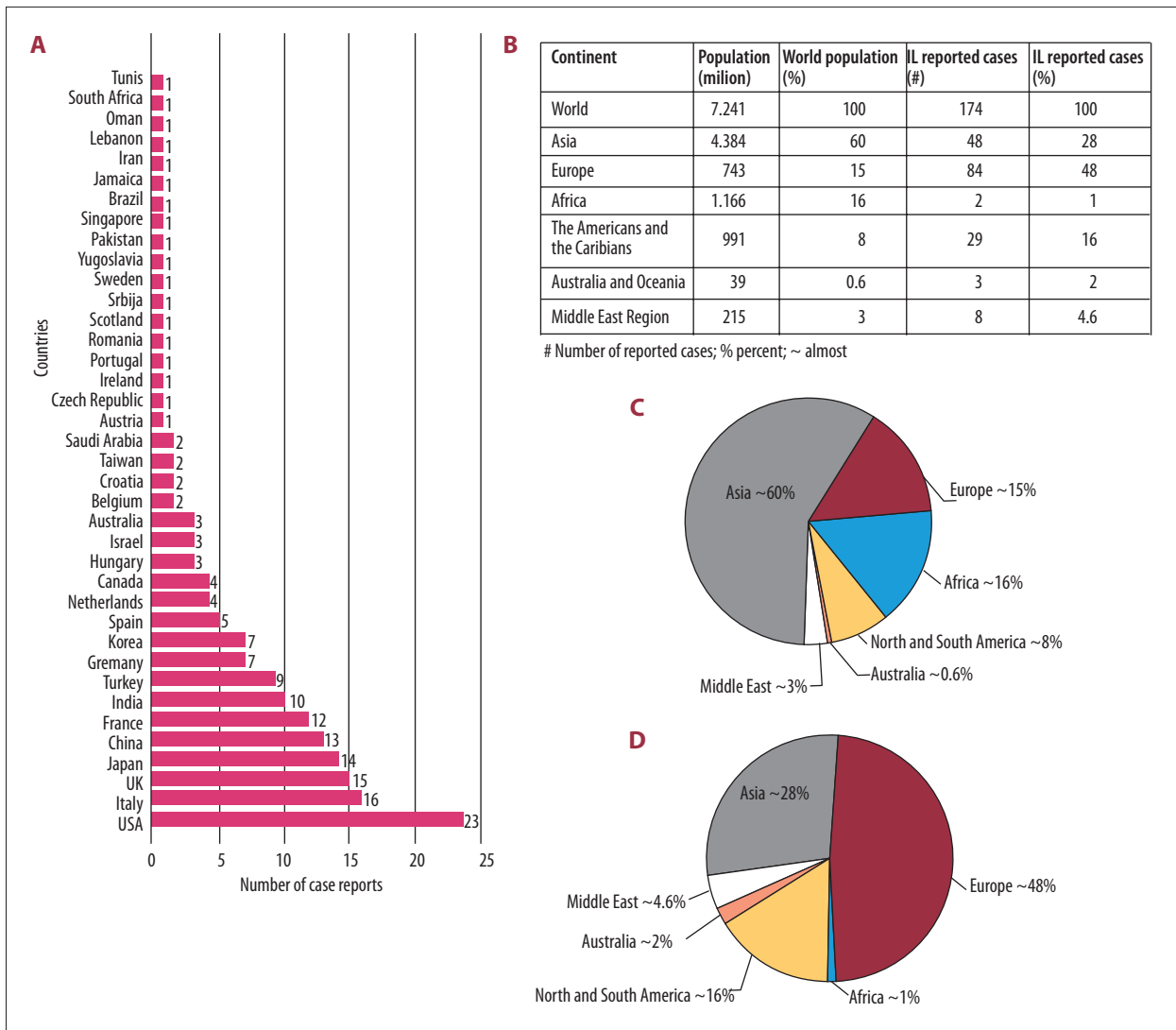


Figure 5. Regional and continental distribution of intestinal lymphangiectasia. The number of reported case reports by the country of disease origin (A); Regional and continental population and the number of reported IL cases (B); Pie chart showing the percent of population in every region and continent (C); Pie chart showing the percent of reported IL cases in every region and continent (D).

We were able to detect the patient's nationality in 174/188 articles. The largest number of reported cases were from Europe: 84/174 case reports (Italy 16, UK 15, France 12, Turkey 9; other European countries had less than five cases per country). We found 48/174 case reports for patients from Asia (Japan 14, China 13, India 10, Korea 7; other Asian countries had less than two cases per country). In North and South America, the United States had the largest number of reported cases, with 23/174. Three cases were reported from Australia, two from Africa and 8/174 were from the Middle East region.

Taking into account the regional and continental population totals, the largest number of reported cases of IL were from countries in Europe, particularly Italy, comprising approximately 50% (Figure 5).

Although in our literature review we cannot point directly to the reason(s) behind the high number of IL cases reported in Europe, on the one hand, we suspect that the medical procedures applied in the countries of the European Union, with regards to health insurance and medical care, covered a more general population, and as a result, discovering and reporting a higher number of rare medical conditions should be expected.

On the other hand, considering the population of Asia in 2015 was almost more than 60% of the world population, and the small number of reported cases from Asian countries, this makes us think that other factors could be the cause of the high number of the reported cases of IL in Europe; naturally, the possibility of genetic factors comes to mind.

Table 2. Medical conditions associated with or complicated by intestinal lymphangiectasia (IL).

PMID*	IL associated with or complicated by	Author	Year
5645698	Nephrotic syndrome	De Sousa JS et al.	1968
4177544	Immunoglobulin A deficiency	Eisner JW et al.	1968
1249692	Noonan syndrome	Herzog DB et al.	1976
604991	Small bowel lymphoma	Ward M et al.	1977
512793	Colonic polyps	Parsons HG et al.	1979
6401772	Macroglobulinemia	Harris M et al.	1983
3984978	Aplasia cutis congenital	Bronspiegel N et al.	1985
3971596	Thymic hypoplasia	Sorensen RU et al.	1985
4018652	Impaired splenic function	Foster PN et al.	1985
4065700	Chylous ascites	Duhra PM et al.	1985
3782656	Pleural effusion	Lester LA et al.	1986
3960333	Vitamin E-deficient spinocerebellar syndrome	Gutmann L et al.	1986
3266226	Epileptic episodes	Katou N et al.	1988
3195550	Systemic sclerosis	van Tilburg AJ et al.	1988
2624276	Facial anomalies and mental retardation	Hennekam RC et al.	1989
2220736	Systemic lupus erythematosus	Edworthy SM et al.	1990
8335455	Constrictive pericarditis	O'Sullivan T et al.	1993
8374252	Acute jejunal ileus	Lenzhofer R et al.	1993
8533807	Zellweger cerebrohepatorenal syndrome	Erdem G et al.	1995
8970209	Oral manifestations	Ralph PM et al.	1996
9932170	Small bowel lymphoma	Gumà J et al.	1998
10403358	Gelatinous transformation of the bone marrow	Marie I et al.	1999
11841381	Xanthomatosis, vaginal lymphorrhoea	Karg E et al.	2002
12124738	Hennekam syndrome	Forzano F et al.	2002
15531848	Incontinentia pigmenti achromians	Riyaz A et al.	2004
16129004	Enamel hypoplasia of the primary dentition	Arrow P et al.	2005
16174162	Breast edema	Goktan C et al.	2005
16407384	Yellow nail syndrome	Danielsson A et al.	2006
16123987	Neuroblastoma	Citak C et al.	2006
17321261	Abdominal mass	Rao R et al.	2007
17514630	Hemolytic uremic syndrome	Kalman S et al.	2007
17354127	Gastrointestinal bleeding	Stovicek J et al.	2007
18431016	Autoimmune polyglandular syndrome type I	Makharia GK et al.	2007
19864203	Vitamin D deficiency	Lu YY et al.	2009
20567835	Castleman's disease	Jeon CJ et al.	2010
21086252	Duodeno-jejunal polyposis	Hirano A et al.	2010
20812055	Digital clubbing	Wiedermann CJ et al.	2010
21947159	Histiocytosis X	Hui CK et al.	2011
22616341	Autoimmune polyglandular syndrome type III	Choudhury BK et al.	2011
22366835	Infantile systemic hyalinosis syndrome	Alreheili K et al.	2012
25276285	Liver fibrosis	Licinio R et al.	2014
26405709	Anemia	Balaban VD et al.	2015
26217101	Generalized warts	Lee SJ et al.	2015

* PMID – PubMed identifier.

In our review we found that a large number of articles (87/188) reported an association between IL and other diseases. Table 2 is a summary of diseases associated with IL, which suggests that a high number of medical cases associated with IL may be due to hypoalbuminemia and lymphopenia and their widespread systemic consequences.

In 37/188 articles, authors introduced diagnostic procedures and radiological findings that can be used to evaluate and diagnose IL, and 15/188 articles shed light on the secondary causes of IL. Table 3 presents a summary of diseases complicated by IL in their clinical course; 30/188 articles described classic cases of IL with all its known clinical features.

Our review found 35/188 articles that described management plans. In particular, octreotide was discussed in 10/35 articles [13–21], antiplasmin in 3/35 [22–24], corticosteroids in 2/35 [25,26], and surgical procedures in 7/35; the rest of the

articles (13/35) described a diet modification plan and the role of MCT oil in the management of IL [27]. In almost all the previous studies, the prognosis was ambiguous due to short time periods of the reported management plans. Octreotide was only effective in blocking diarrheal episodes and for correcting hypoalbuminemic states. Table 4 presents a summary of studies that discussed the role of octreotide in the management of IL.

In our patient's case, after more than three years of a careful management plan and persistent follow-up, it was clear that octreotide, diet modification, and MCT oils were unable to eliminate the edema. Using MCT oils over more than three years did not show any important improvement in the edema size (Figure 6). However, these management approaches were of benefit in restricting further extension of the edema, although they were unable to eradicate the existence of the original presenting edema. Most likely, this is related to the fact that MCT oils and octreotide are unable to affect already-extended

Table 3. Secondary causes for IL.

PMID	Disease	Author	Year
7463809	Myxedema heart	Munakata Y et.al	1980
7247748	Systemic lupus erythematosus	Pereira AS et.al	1980
3608736	Radiotherapy and chemotherapy	Rao SS et.al	1987
17632262	Waldenstrom macroglobulinemia	Pratz KW et.al	2007
20801422	Primary peritoneal carcinoma	Steines JC et.al	2010
21092952	Multiple myeloma	Bhat M et.al	2011
21837942	Primary hypoparathyroidism	Koçak G et.al	2011
23591283	Diffuse large B-cell lymphoma	Patel KV et.al	2013

Table 4. Octreotide management in IL.

PMID	Age	Gender	Indication	Outcome	Author	Year
7783555	38	M	Hypoalbuminemia	Improvement#	Bac DJ et al.	1995
11227670	21	M	Hypoalbuminemia	Improvement	Kuroiwa G et al.	2001
12924644	27	F	Generalized hydrops	May be effective	Klingenberg RD et al.	2003
15506669	25	F	Severe edema and diarrhea	Improvement	Filik L et al.	2004
15565209*	–	–	Severe edema	No improvement	Makhija S et al.	2004
15373983	–	–	Edema, diarrhea	Improvement	Balboa A et al.	2004
21768882	15	M	Hypoalbuminemia	Improvement	Altit G et al.	2012
23555496	63	M	Edema, diarrhea	Improvement	Suehiro K et al.	2012
23180957	Baby	–	Edema	Improvement	Al Sinani S et al.	2012

Improvement: Correction of hypoalbuminemia; * octreotide was tested to treat intestinal lymphangiectasia in guinea pig model.

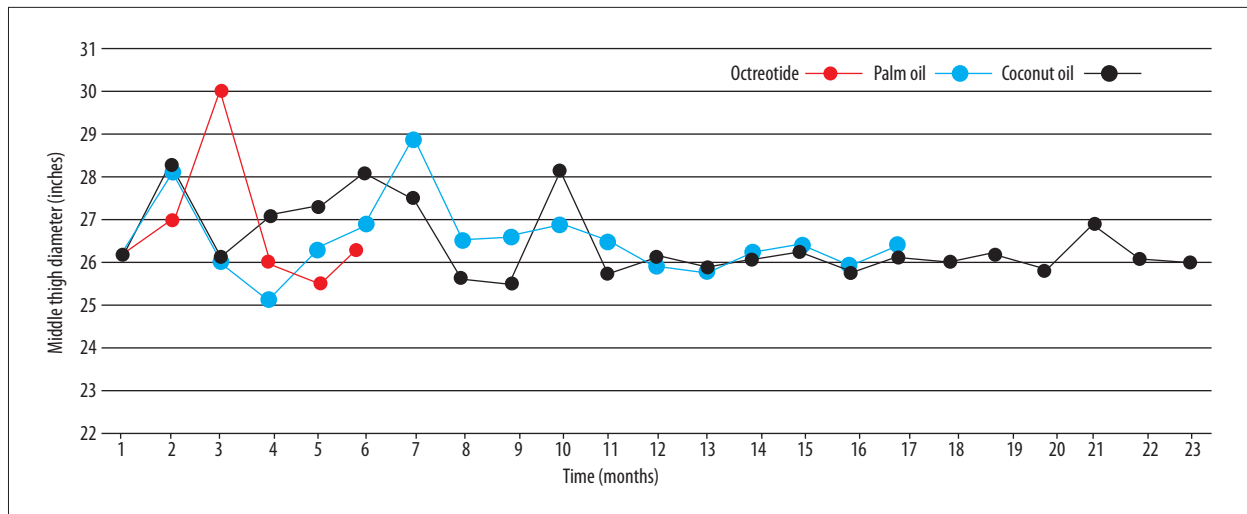


Figure 6. The changes observed in edema size under management over time. Line chart showing the change in edema size over time following management with octreotide (red); palm oil (blue); coconut oil (black). The treatments were not applied simultaneously, but overlapped for easy interpretation of the data.

lymphatic vessels. Notably, the only benefit observed for the use of MCT oils is that it, probably, played a role in maintaining the immune system function against various infectious agents. This is in part due to the fact that these oils can help in preventing the loss of enteric proteins.

Conclusions

Low-fat diet plans, light exercises, massages, and medium-chain triglycerides (MCT), such as coconut and palm oils, can help to prevent further extension of edema in patients with (IL). It seems that MCT oils are an excellent choice to protect

against further loss of body proteins, and this can help to boost the immune system against various infectious agents. It is very important to note that octreotide and MCT oils had no effect on reducing lymphedema in our patient.

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Statements and declarations regarding conflicts of interest

The authors have no conflicts of interest to disclose.

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