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Case report

Congenital intestinal lymphangiectasia. A case report

Georgios Mitsiakos[1], Eftychia Drogouti[1], Maria Drogouti[2], Charalampos Doitsidis[3], Elisabeth Pazarli[4], Ioannis Spyridakis[3]

[1]Second Department of Neonatology and Neonatal Intensive Care Unit, Aristotle University of Thessaloniki, "Papageorgiou" Hospital, Thessaloniki, Greece

[2]Department of Surgery, Agios Dimitrios General Hospital, Thessaloniki, Greece

[3]Second Department of Pediatric Surgery, Aristotle University of Thessaloniki, "Papageorgiou" Hospital, Thessaloniki, Greece

[4]Anatomical Pathology Laboratory, "Papageorgiou" Hospital, Thessaloniki, Greece

Abstract

Congenital intestinal lymphangiectasia, first described by Waldmann et al. in 1961, is a rare congenital malformation of the lymphatics, presented with generalized edema, hypoproteinemia and lymphopenia. Diagnosis is based on endoscopy findings and pathology.

We present here a case of a male neonate, second child of an indigenous woman, delivered by caesarean section. Prenatally, multiple cystic abdominal masses were identified by ultrasound. The patient was treated successfully with enterectomy and anastomosis. Histopathology revealed primary intestinal lymphangiectasia with no features of malignancy.

Intestinal lymphangiectasia is a rare pathology, which should be differentiated while exploring abdominal masses, hypoproteinemia and edema especially in neonates.

Keywords

Congenital intestinal lymphangiectasia, neonatal abdominal masses, lymphatics disorders.

Corresponding author

Georgios Mitsiakos, Second Department of Neonatology and Neonatal Intensive Care Unit, Aristotle University of Thessaloniki, "Papageorgiou" Hospital, Thessaloniki, Greece; email: mitsiakos@auth.gr.

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Introduction

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 and immunological abnormalities [1]. It may occur as a congenital disorder or be acquired due to lymphoma, retroperitoneal masses, Crohn's disease or mesenteric tuberculosis.

Congenital intestinal lymphangiectasia (CIL) was originally described by Waldmann et al., in 18 patients presenting with edema and hypoproteinemia caused by dilated lymph vessels of the intestine [2]. The prevalence of clinically overt CIL is unknown [3]. Edema is the main clinical presentation. Other clinical signs include ascites, pleural effusion and pericarditis [3]. Hallmark biochemical characteristics include lymphopenia, hypoalbuminemia and hypogammaglobulinemia, as well as protein-losing enteropathy [4].

Diagnosis is based on characteristic findings during the double-balloon enteroscopy and their review from a pathology perspective. Medical options include dietary modifications, octreotide, tranexemic acid and vitamin D supplementation. Small bowel resection is useful in the rare cases where intestinal lymphangiectasia is segmental and localized [3].

Case report

A two-day male neonate was admitted to the Pediatric Surgery Department on referral from a general hospital in order to assess and treat an abdominal mass prenatally diagnosed by ultrasound (**Fig. 1**). The neonate was delivered by caesarian section at the gestational age of 37 weeks due to mother's hypertension and a prior caesarean section. The patient was the second child of an indigenous family. Maternal history included hypertension. Both parents were free from family history consistent to CIL. Apgar score was 8 and 9 at 1 and 5 minutes, respectively, and birth weight was 2,240 g. Clinical examination of the neonate on admission was normal and a laboratory work-up proved hypoalbuminemia. Abdominal ultrasound scan on first and third day of life revealed a multilocular cystic mass of 7 x 7 x 3 cm dimensions. Abdominal MRI identified multiple cystic lesions of the mesentery with features compatible with lymphangiomas.

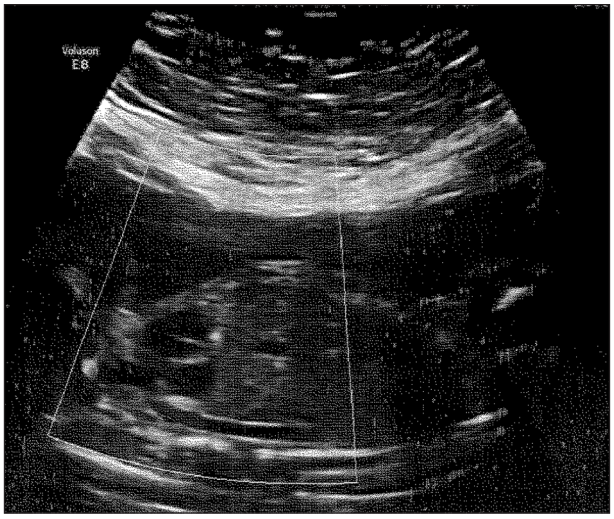

Figure 1. Prenatal ultrasound findings. Multiple cystic lesions of the mesentery.

The neonate was scheduled for laparotomy on the third day of life. Median laparotomy was performed, disclosing multiple cystic lesions of the mesentery between jejunum and ileum (**Fig. 2** and **Fig. 3**). Duodenum, colon, liver and spleen were normal. A wide excision of the involved mesentery and adjacent small intestine was performed with a hand-sewn end-to-end anastomosis between the proximal and the distal end of the small intestine. The surgical procedure was uneventful and a drain was placed in the abdominal cavity, near to the anastomosis. The neonate was admitted in the Neonatal Intensive Care Unit for post-operative care. Extubation was performed on the 1st post-operative day (POD). On the 2nd POD the neonate was discharged to the Pediatric Surgery Department on total parenteral nutrition and triple antibiotic treatment. He resumed oral intake on POD 5. The drainage catheter was removed on POD 6. Hospitalization was uneventful and the patient was discharged home on POD 18 with diet instructions.

Histopathology of the lesions revealed multiple dilated lymph vessels along the intestinal wall; such findings were compatible with CIL (**Fig. 4** and **Fig. 5**). No features of malignancy were marked. No adjuvant treatment was applied and no local recurrence was found during the following month.

Discussion

IL is a rare condition affecting the intestinal lymph vessels and obstructing the lymphatic drainage of the intestine [5] due to primary ectasia

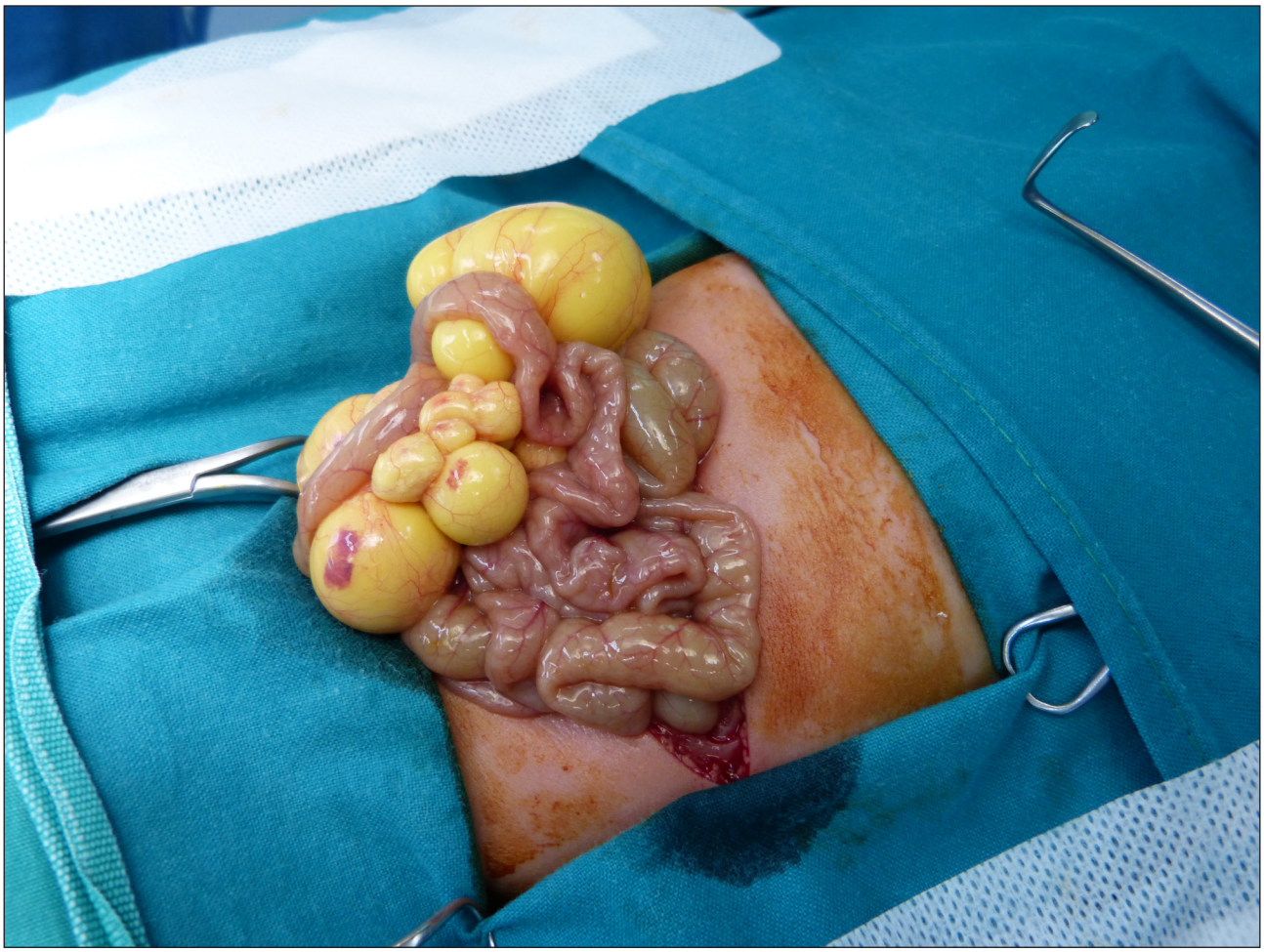

Figure 2. Multiple cystic lesions of the mesentery between jejunum and ileus.

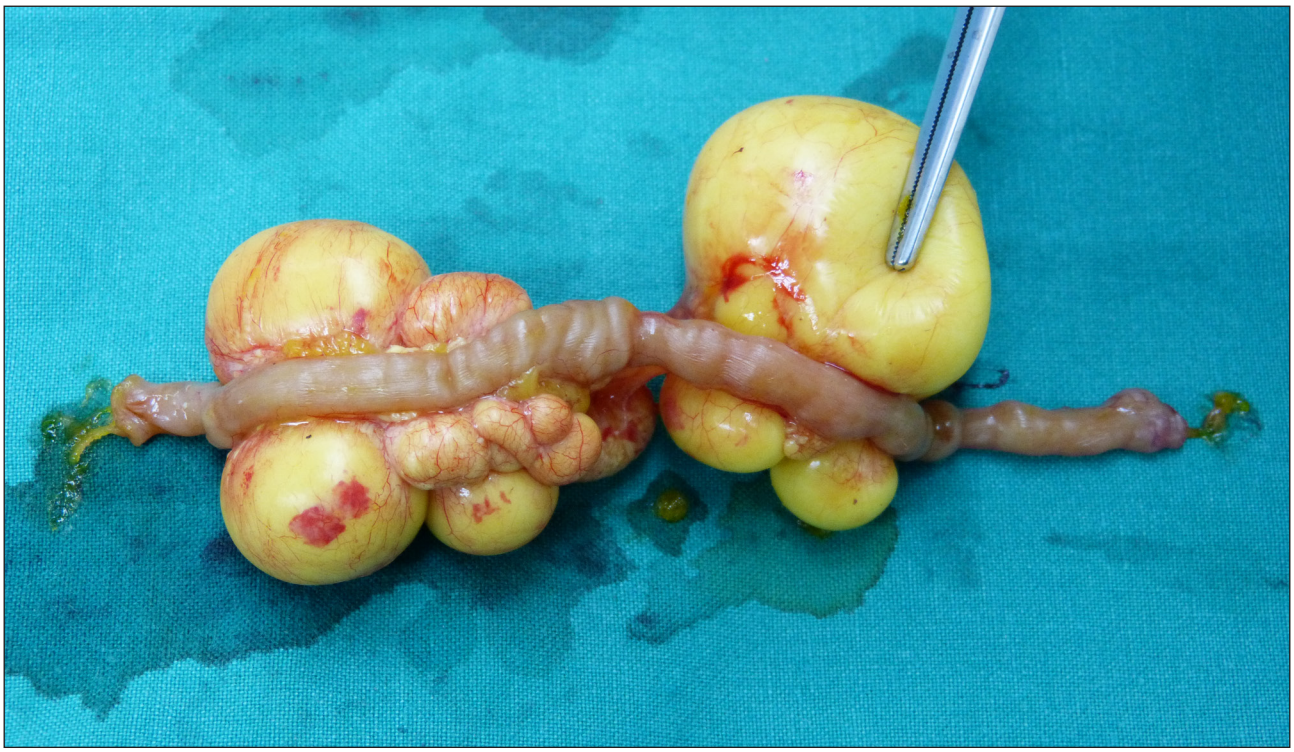

Figure 3. Fragment of small intestine with multiple cysts containing serous-white fluid.

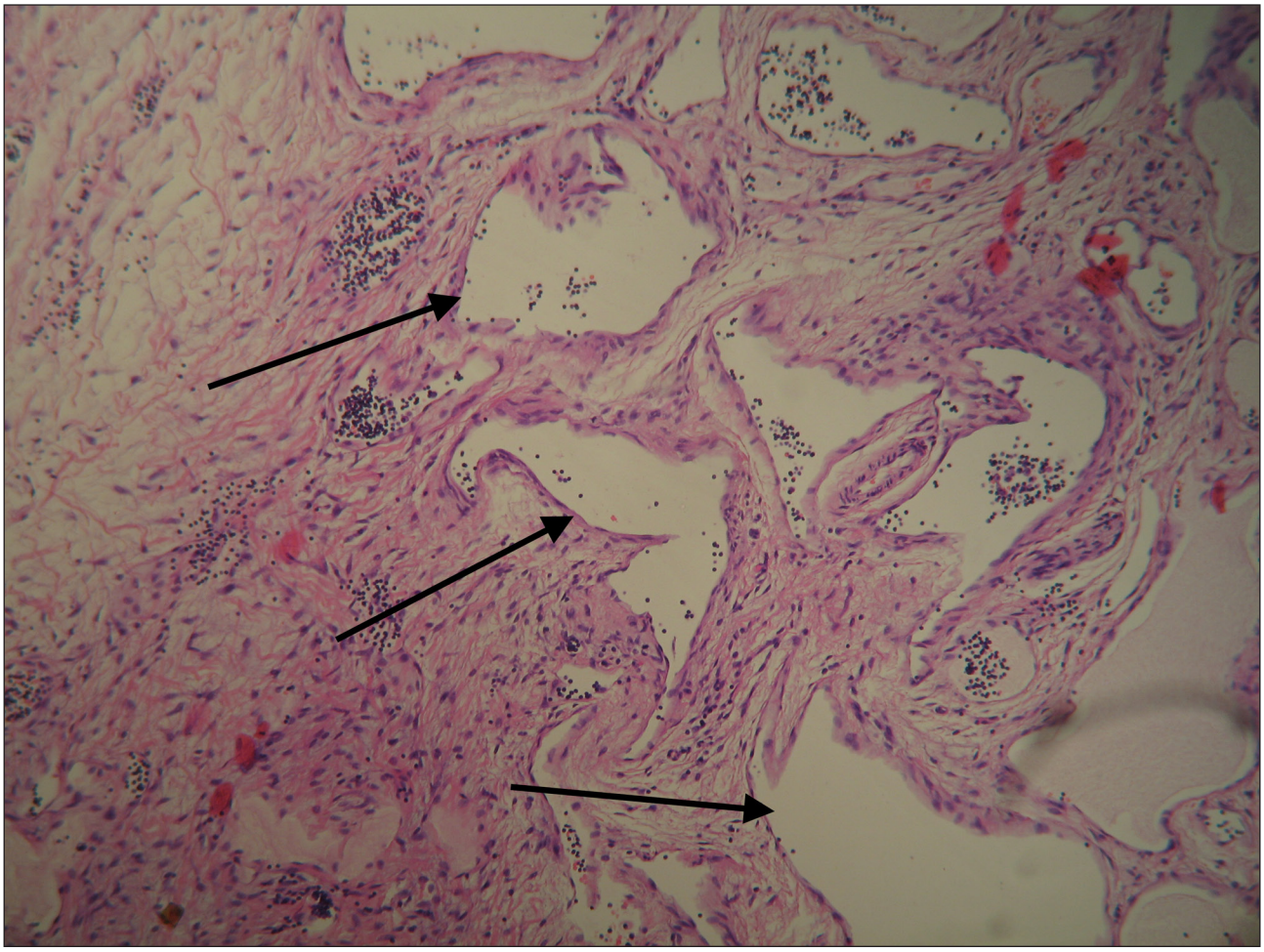

Figure 4. Dilated lymphatics located in the intestinal wall (AE x 200).

or as a result of secondary impacts on the normal lymph circulation, such as heart diseases and retroperitoneal lymph node enlargements [1]. CIL is an unusual cause of protein losing enteropathy either due to congenital malformation or obstruction of intestinal lymphatics [2]. The etiology of CIL still remains unknown [6-11]. It presents with edema of variable degree and childhood particularities such as inability to gain weight and growth retardation. Other clinical sings may include lymphedema, abdominal mass, malabsorption syndrome, mechanical ileus, chylous reflux, celiac disease, necrolytic migratory erythema, recurrent hemolytic uremic syndrome and osteomalacia [3]. Laboratory findings include decreased albumin and total protein levels [2]. Loss of lymphatic fluid-containing lymphocytes in the intestine is also a specific finding and loss of CD4+ T cells is predominantly observed [7]. Interestingly, in this present case the only laboratory abnormal finding was hypoproteinemia and clinical examination revealed no pathological clinical signs such as ascites or edema, possibly due to early detection and treatment. CIL is usually diagnosed earlier than the third year of age, although diagnosis may occur later on, in late childhood [6]. The diagnosis of CIL is difficult, because abnormal lymphatic lesions are usually distributed in the small intestine, as were in our patient. Detection of lesions by upper gastroscopy and colonoscopy is limited. As a result, double balloon enteroscopy or surgical methods are used for pathological examination in some cases [7]. Endoscopy was not the diagnostic method of choice in our neonate because MRI scan revealed lesions compatible with lymphangioma and thus surgery was preferred. Diagnosis of intestinal lymphangiectasia is established by the characteristic histology of grossly dilated lymphatics seen in the lamina propria of the small bowel (duodenum/jejunum/ileum) [11]. Another test proposed is albumin scintigraphy, which shows the albumin leakage into the bowel [6]. Given the preoperative data in this present case (segmental cystic masses initially compatible with lymphangioma), surgery was the method of choice for both diagnosis and treatment, and definitive diagnosis was based on histology of the surgical specimen. Microscopically

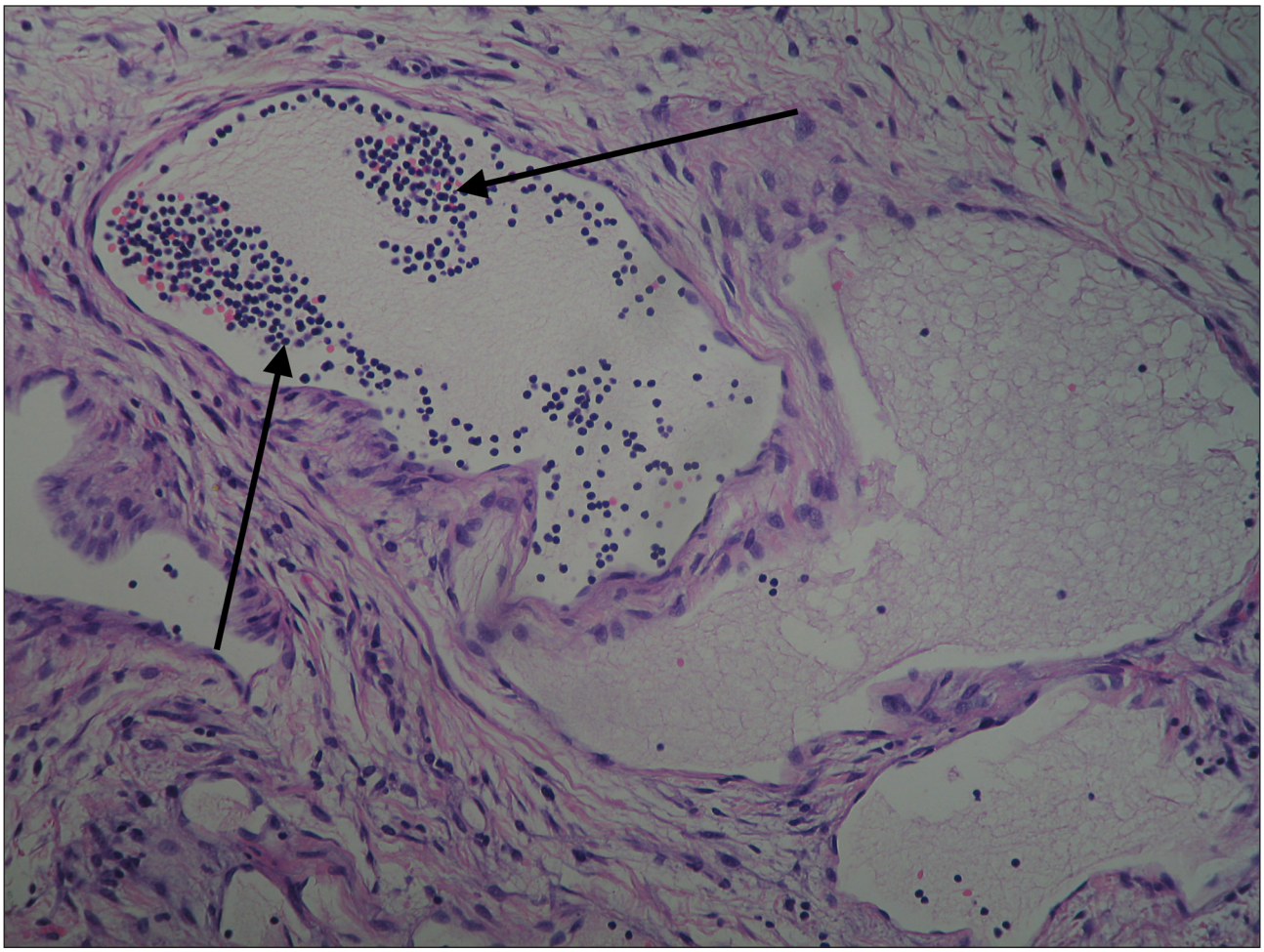

Figure 5. Intraluminal lymphocytes within dilated lymphatics of the intestinal wall (AE x 400).

the intestinal specimen presented multiple cystic lymph vessels along the intestinal wall, the hallmark lesion of the disease.

Treatment of CIL depends on the severity and extent of involvement. For most patients with CIL, due to generalized abnormalities and diffuse distribution, dietary modification with a low-fat, high-protein diet and supplementation of medium-chain triglycerides (MCT) is the mainstay of treatment [9]. Segmental resection may be recommended in cases involving localized segment of the intestine [10], as concerned operative findings in our case, and can successfully treat protein losing enteropathy, anemia or abdominal pain [9]. The clinical outcome in our patient was excellent, as the lesions were segmental and sufficient length of small intestine was preserved. No dietary modifications were required given the extent of the disease.

Conclusion

We report a rare case of CIL incidentally diagnosed on prenatal control. Diagnosis was established on the pathology findings after the surgical removal of an abdominal mass caused by CIL. Surgery was performed on the third day of life. Hypoproteinemia was the only abnormal laboratory finding. Due to early diagnosis and treatment, enteropathy, edema or ascites did not occur.

CIL is a rare entity, which should be differentiated while exploring hypoproteinemia and/or abdominal masses especially in children and infants. Treatment of CIL depends on the extent of the involvement. Surgery may be curative when lesions are segmental.

Declaration of interest

The Authors declare that there is no conflict of interest.

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