A Rare Case of Primary Intestinal Lymphangiectasia Presenting as Recurrent Hypocalcemic Seizures

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Abstract - Background: Primary Intestinal Lymphangiectasia (PIL) is a rare disorder characterized by protein-losing enteropathy due to dilated intestinal lymphatics, leading to hypoalbuminemia, lymphopenia, and malabsorption. Its presentation with recurrent hypocalcemic seizures is highly unusual.

Case Presentation: We report the case of a 6-month-old male who presented with recurrent generalized tonic-clonic seizures, preceded by episodes of non-bloody diarrhea. Initial workup revealed severe hypocalcemia, hypoalbuminemia, and hypomagnesemia. Infective and renal causes were ruled out. The infant had a history of inadequate nutrition, compounded by maternal postpartum depression and cholecystectomy. Despite calcium, magnesium, and vitamin D supplementation, symptoms recurred, necessitating further evaluation.

Endoscopic biopsy revealed dilated, globular, whitish villi in the terminal ileum, confirming the diagnosis of Primary Intestinal Lymphangiectasia. The child was managed with albumin infusion, vitamin supplementation, and subcutaneous octreotide. A dietitian-recommended high-protein, fat-restricted, MCT-based diet was initiated, leading to clinical improvement.

Conclusion: This case highlights an uncommon presentation of PIL with recurrent hypocalcemic seizures, emphasizing the importance of considering protein-losing enteropathy in cases of unexplained hypocalcemia, hypoalbuminemia, and malabsorption. Early recognition and dietary modifications are crucial for long-term management.

INTRODUCTION

Primary Intestinal Lymphangiectasia (PIL) is a rare protein-losing enteropathy caused by congenital malformations of the intestinal lymphatics. It typically presents with peripheral edema, diarrhea, and serous effusions, but neurological manifestations due to electrolyte imbalances are rarely reported.

CASE PRESENTATION

A 6-month-old male presented with three episodes of generalized tonic-clonic seizures following 4–5 episodes of greenish, watery diarrhea. There was no fever, abdominal pain, or rash. The child was born at term via LSCS, with an unremarkable antenatal and birth history. The mother had undergone a postpartum cholecystectomy and had postpartum depression, leading to inadequate formula feeding. Neither the infant nor the mother received vitamin D or calcium

INVESTIGATIONS

On evaluation, the child had severe hypocalcemia, hypomagnesemia, and hypoalbuminemia. Infectious and renal causes were ruled out. Initially managed with intravenous and oral calcium, magnesium sulfate, and vitamin D3, the child improved but relapsed a month later with similar symptoms. Refractory hypocalcemic seizures raised suspicion of an underlying malabsorptive disorder.Endoscopy and biopsy confirmed PIL, revealing dilated, globular, whitish villi in the terminal ileum.

TREATMENT

The child was treated with albumin infusion, vitamin K, D, calcium, and magnesium supplementation. Subcutaneous octreotide was administered for a month, and a high-protein, fat-restricted MCT-based diet was initiated, leading to clinical stability. The mother was also found to have hypocalcemia and vitamin D deficiency, requiring supplementation.

DISCUSSION

PIL is rare, and its exact incidence is unknown. It may be primary due to congenital lymphatic malformations or secondary to inflammatory disorders like Crohn's disease, tuberculosis, or lymphoma. It typically presents with hypoalbuminemia-induced edema, but in this case, recurrent hypocalcemic seizures were the initial manifestation.

The mainstay of treatment includes dietary modifications, particularly a low-fat, MCT-based diet. Octreotide has shown promise in improving biochemical and histological parameters, though its exact mechanism remains unclear.

CONCLUSION

This case underscores the importance of considering PIL in infants with unexplained recurrent hypocalcemia and malabsorption. Early diagnosis and dietary intervention play a crucial role in management, improving prognosis and quality of life.

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