

CASE REPORT

TENSION PNEUMOPERITONEUM: A RARE PRESENTATION OF MEGACYSTIS MICROCOLON INTESTINAL HYPOPERISTALSIS SYNDROME

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Megacystis microcolon intestinal hypoperistalsis syndrome also known as Berdon syndrome is characterized by enlarged urinary bladder, small colon and reduced or absent intestinal peristalsis. We report a case of 4 days old female suffering from MMIHS presenting with tension pneumoperitoneum. To the best of our knowledge, this is the first reported case of MMIHS, having this unusual presentation.

Keywords: Tension pneumoperitoneum; Megacystic microcolon; Intestinal hypoperistalsis; syndrome; Perforation; Intestinal malrotation

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INTRODUCTION

Megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) was first described by Berdon *et al* in 1976.¹ It has an autosomal recessive pattern of inheritance, having preponderance for female gender.² Antenatal and post-natal imaging is the mainstay of diagnosis. Cardinal features are distended urinary bladder, bilateral hydronephrosis and polyhydramnios.³ Treatment is supportive and prognosis is usually poor with short survival.

CASE REPORT



Figure-1: Tension pneumoperitoneum centrally displacing the abdominal viscera on radiograph erect abdomen.

The patient was the first-born female child of consanguineous parents, delivered by caesarean

section at 34 weeks of gestation. Three days after birth she presented to emergency department with increasing abdominal distension. Her antenatal ultrasound was reviewed that revealed a markedly distended urinary bladder occupying most of the foetal abdomen with moderate bilateral hydronephrosis and polyhydramnios. Abdominal radiograph showed tension pneumoperitoneum which was decompressed followed by emergency exploratory laparotomy. The per-operative findings showed enlarged urinary bladder, intestinal malrotation, ladd bands, sigmoid perforation and microcolon. Her vesicostomy and colostomy was performed. Supportive treatment and total parenteral nutrition was administered in neonatal ICU. Her family was counselled regarding her disease outcome. She stayed in hospital for 14 days and was shifted to a public sector hospital on her parent's request, where she died at the age of 25 days.

DISCUSSION

Megacystis microcolon intestinal hypoperistalsis syndrome is a neonatal intestinal motility disorder.¹ It usually presents with massive abdominal distension, hypoperistalsis and bilious vomiting. However, our case is unique in its presentation with tension pneumoperitoneum secondary to sigmoid perforation. The typical operative findings include enlarged urinary bladder, microcolon and malrotation of the small intestine which corresponds to our case as well. Literature review suggests multiple factors contributing towards its pathogenesis like degenerative disease of smooth muscle, innervation defects, in-utero inflammation of urinary and gastrointestinal tract and drugs.⁴ Antenatal ultrasound of our patient revealed grossly distended urinary bladder and bilateral moderate hydronephroureter

which is a reliable sign for the diagnosis of MMIHS. The important differentials include prune belly syndrome, bowel atresia, volvulus and Hirschprung's disease. Presence of non-obstructed urinary bladder in MMIHS and normal or increased amniotic fluid volume differentiates it from prune belly syndrome which occurs almost exclusively in males.⁴ Nutritional support and palliative surgery remain the mainstay of management.⁵

CONCLUSION

MMIHS is a rare disorder with grim outcome. Sound knowledge regarding imaging features of this disease aids in antenatal diagnosis. Enlarged urinary bladder on antenatal ultrasound and polyhydramnios should raise the suspicion of MMIHS. Moreover, in settings of neonatal pneumoperitoneum and gut perforation, MMIHS should be considered as a likely differential and managed accordingly.

REFERENCES

1. Berdon WE, Baker DH, Blanc WA, Gay B, Santuli TV, Donovan C. Megacystis-microcolon-intestinal hypoperistalsis syndrome: a new cause of intestinal obstruction in the newborn - Report of radiological findings in five newborn girls. *AM J Roentgenol* 1976;126(5):957-64.
2. Anneren G, Meurling S, Olsen L. Megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS): an autosomal recessive disorder - Clinical reports and review of the literature. *Am J Med Genet* 1991;41(2):251-4.
3. Verbruggen SC, Wijnen RM, van den Berg P. Megacystis microcolon-intestinal hypoperistalsis syndrome: a case report. *J Matern Fetal Neonatal Med* 2004;16(2):140-1.
4. Manop J, Chamnanvanakij S, Wattanasarn C. Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS): A Case Report in Thailand. *J Med Assoc Thai* 2004;87(11):1385-8.
5. Puri P, Lake BD, Gorman F, O'Donnel B, Nixon HH. Megacystis-microcolon-intestinal hypoperistalsis syndrome: a visceral myopathy. *J Pediatr Surg* 1983;18(1):64-9.

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