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Allogeneic Transplant

Intestinal pneumatosis in the context of Hematopoietic Stem Cell Transplantation (HSCT): a case series

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Pneumatosis intestinalis (PI) is a rare disorder characterized by extensive gas collections in the intestinal submucosa that can develop into abdominal rupture. The diagnosis is established based on imaging studies such as computed tomography (CT). In post-HSCT patients, their origin can be multifactorial, such as conditioning, infections, graft-versus-host disease (GVHD) and prolonged use of steroidal anti-inflammatory drugs (SAIDs). Objective: To report 4 cases of PI and discuss the presentation and management in the context of HSCT. Case 1: 23-year-old female patient, with Myelodysplastic Syndrome, submitted to related 10x10 allogeneic HSCT. After 6 months, the patient developed watery diarrhea and weight loss, with acute GVHD in the GIT being confirmed by duodenal biopsy. She received methylprednisolone 2 mg/ kg/ day and oral budesonide, with no improvement. He received 2 infusions of basiliximab, with partial improvement. After 30 days, she suddenly developed painful abdominal distension and dyspnea. CT scan showed PI with pneumoperitoneum and pneumomediastinum. She developed septic shock (SS) and started receiving broad antimicrobial therapy (AMT), zero diet by mouth (NPO) and parenteral nutrition (TPN). After 15 days, she partially reabsorbed the pneumatosis, but died due to candidemia by *C. tropicalis*. Case 2: 8-year-old male patient old, with Wiskott-Aldrich syndrome, submitted to the 2nd 10x10 unrelated (UNR) allogeneic HSCT due to loss of the previous graft. He developed acute skin and GIT GVHD with good response to SAIDs. After two years, he developed abdominal pain and fever, and the CT scan showed PI. He required AMT, NPO and TPN for prolonged periods. He showed complete resolution of the condition. Case 3: One-year-old female patient, submitted to UNR 10x10 HSCT due to Combined Immunodeficiency Syndrome. After 90 days she started with diarrhea, bloating and weight loss. She was diagnosed with GIT GVHD, treated with SAIDs, with partial improvement. However, she developed SS and the CT showed PI. She received AMT, NPO and TPN, with clinical picture resolution in three weeks. After one month she had a new episode of SS and died. Case 4: A 12-year-old female patient was submitted to UNR 9x10 HSCT for acute lymphoblastic leukemia. After the engraftment she had acute skin GVHD with good response to SAIDs. On D+164, she started to have fever and malaise. CT showed extensive PI and pneumoperitoneum. The patient remained stable, treated with AMT and NPO. She did not need TPN. She progressed well and was discharged after 4 weeks. Discussion: Pl is a rare late complication of HSCT with high morbidity and mortality. We demonstrated 4 cases of PI, of which onset occurred after the first 100 days with concomitant diagnosis of GVHD of TGI and use of SAIDs. Conclusion: Treatment of PI remains controversial and conservative or surgical management can be performed. Our patients showed good evolution with GIT rest, TPN and broad spectrum AMT. Nevertheless 3 of them died, but only during the first episode of PI, which may represent mortality that is not directly related, but a marker of severity in HSCT.

Keywords: Transplantation, Stem Cells, Pneumatosis, GVHD, Antimicrobials