

# Pseudomonal Perianal Necrotizing Soft-Tissue Infection in a Pediatric Patient with Hodgkin Lymphoma

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<b>Background</b>	The hospitalization of an eleven-year-old female with Hodgkin lymphoma was complicated by a pseudomonal necrotizing soft-tissue infection of the anus.
<b>Summary</b>	An 11-year-old female presents with nausea, vomiting, diarrhea, and anorexia six days after initiating her third cycle of chemotherapy for stage IVa lymphocyte predominant Hodgkin lymphoma. She had recently completed a ten-day course of amoxicillin/clavulanate for bilateral onychocryptosis one week prior to starting her chemotherapy. Vincristine, bleomycin, and prednisone were given per chemotherapeutic protocol. Upon evaluation, her stools were positive for <i>Clostridium difficile</i> , and metronidazole was started. Her course was complicated by pancytopenia, coagulopathy, and <i>Pseudomonas</i> bacteremia. She then began complaining of perianal pain. An MRI revealed significant proctitis with concern for perianal fistula. She was found to have a perianal necrotizing soft tissue infection, necessitating serial debridement and eventual diverting colostomy. Tissue cultures demonstrated growth of <i>Pseudomonas aeruginosa</i> ( <i>P. aeruginosa</i> ). While the increased risk of perianal sepsis in neutropenic patients is well documented, the progression to necrotizing soft tissue infection is far less common in the literature. This case report demonstrates a rarely encountered, but often lethal, complication of hematologic malignancies and chemotherapy that can be difficult to detect clinically. The need for high clinical suspicion and early surgical consultation is emphasized by this case report.
<b>Conclusion</b>	Neutropenia associated with hematologic malignancy is a known risk factor for development of perianal sepsis. We present the case of a pseudomonal perianal necrotizing soft tissue infection in an eleven-year-old female undergoing chemotherapy for Hodgkin lymphoma. This case highlights the need for high clinical suspicion and an early multidisciplinary approach in this population.
<b>Keywords</b>	Perianal necrotizing soft tissue infection, perianal sepsis, hematologic malignancy, Hodgkin lymphoma, <i>Clostridium difficile</i> , <i>Pseudomonas aeruginosa</i>

**DISCLOSURE:**

The authors have no conflicts of interest to disclose.

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## Case Description

Perianal sepsis in patients with hematologic malignancy is a well-established phenomenon.<sup>1-6</sup> This arises secondary to immunosuppression caused both by the malignant process and the treatment regimen.<sup>5</sup> The presentation of these patients is atypical when compared to the healthy patient, with non-specific symptoms such as fever and pain often being the only clinical clues to the developing infection.<sup>1-2,4,7-9</sup> We submit a unique presentation of the rapid and devastating course that these infections can take.

The patient is an eleven-year-old female with stage IVa lymphocyte predominant Hodgkin lymphoma who was being treated with a chemotherapy regimen consisting of bleomycin, etoposide, doxorubicin, cyclophosphamide, vincristine, procarbazine, and prednisone (BEACOPP). Eight days prior to starting her last round of chemotherapy, the patient completed a ten-day course of amoxicillin/clavulanate for onychocryptosis. She had diarrhea associated with this treatment, and this resulted in perianal discomfort attributed to hemorrhoids. These symptoms had abated prior to arrival for chemotherapy. Per the oncology team, she received two days of clindamycin at the start of her chemotherapy for onychocryptosis. She began cycle three of three and was subsequently readmitted to the hospital seven days later with diarrhea, nausea, vomiting, anorexia, and fatigue; these symptoms had begun one day prior to admission. Upon admission for her diarrhea, her stools were positive for *Clostridium difficile*, and she was started on oral metronidazole. The patient began complaining of escalating perianal pain on hospital day two. This was initially attributed to recrudescence of her hemorrhoidal discomfort. On hospital day three, she developed sepsis, and blood cultures were positive for *Pseudomonas aeruginosa* (*P. aeruginosa*). Piperacillin-tazobactam and gentamicin were added.

The patient's perianal pain became uncontrolled, prompting magnetic resonance imaging (MRI) and surgical consultation on hospital day four. MRI results demonstrated evidence of proctitis with a right posterior anocutaneous fistula without associated abscess. Her physical examination was difficult given her age and discomfort. However, exam demonstrated external hemorrhoids with thromboses, granulation tissue without erythema or active drainage, edema, and excoriation of the perineal body. Recommendation at that time was for continuation of systemic antibiotics, sitz baths, and zinc oxide ointment, with possible exam under anesthesia if symptoms remained concerning. The patient's symptoms improved on hospital day five,

allowing continuation of conservative management. However, because of persistence of pain, exam under anesthesia was performed on hospital day six. This demonstrated significant anal and perianal necrosis with sparing of the rectal mucosa. She underwent wide debridement of the necrotic tissue, which included significant portions of her external sphincter and partial excision of her internal sphincter. Tissue specimens were sent to pathology which demonstrated "skin, underlying fibroadipose, skeletal muscle, and smooth muscle with necrosis and areas of perivascular and intravascular organisms appearing to be bacteria, some gram-positive cocci, and many variable gram-negative rod/filamentous forms." This was consistent with her diagnosis of necrotizing fasciitis, with additional findings suggestive of blood-borne source of infection, including organisms within the walls of the blood vessels.

Over the following weeks, the patient underwent six return trips to the operating room, ultimately debriding 85 percent of her external sphincter and portions of her internal sphincter and rectovaginal septum, leaving the puborectalis and majority of internal sphincter intact. As more tissue needed to be debrided and the wound could no longer be controlled by a fecal management system, the need for fecal diversion became clear and she underwent laparoscopic diverting end sigmoidostomy ten days after her initial operation. The perianal wound was then able to be managed by negative pressure therapy. Eighteen days following her initial surgery, the remaining tissue was healthy, and she underwent repair of rectovaginal fistula, rectal sleeve advancement, and split-thickness skin grafting over her perianal wound (Figure 1 and Figure 2).



**Figure 1.** Completed serial debridements with exposed ischioirectal adipose tissue and minimal remaining anal canal.



**Figure 2.** Perianal wound following split-thickness skin grafting.

The skin graft had greater than 95 percent take with excellent approximation of the rectal mucosa to the graft. She was discharged on hospital day 43. She completed radiation treatment to her primary right groin site, with limited scatter to the perianal area as the final oncologic treatment for her primary Hodgkin lymphoma. At five months after her rectal sleeve advancement, anorectal manometry was performed, which demonstrated no resting anal tone nor any voluntary squeeze pressure as well as rectal hypersensitivity with initial urge at 20 milliliters and intense urge at 40 milliliters. The patient has undergone pelvic floor physical therapy using an internal sensor with minimal clinical improvement in squeeze capability. The family wishes to pursue a trial of colostomy takedown with consideration of continence adjuncts including thickening of stool, injectable bulking agents, and sacral nerve stimulator. Potential need for repeat colostomy has been discussed.

The patient's care team has included medical oncology, radiation oncology, general and colorectal surgery, plastic surgery, infectious disease, pain and palliative care, physical therapy, occupational therapy, psychology, psychiatry, wound and ostomy nursing, and oncology nursing, as well as countless others participating in her care. She will con-

tinue to have input from a number of sources in both the pediatric, as well as the adult realm, as we move forward with her care.

## Discussion

Perianal sepsis, defined as the bacterial invasion of the soft tissues in the perianal area with associated systemic responses, is a known complication in patients with hematologic malignancies, with prevalence ranging between two to nine percent.<sup>1-6</sup> It is also described as having an increased association with an underlying disorder in pediatric patients, such as inflammatory processes or neoplasms, when compared to perianal sepsis in adults.<sup>10</sup> However, what is far less commonly described in the literature is the devastating end of this disease spectrum, specifically perianal necrotizing soft tissue infections. These necrotizing infections are rare but highly fatal, with an early study demonstrating 100 percent mortality<sup>11</sup> and a more recent study demonstrating a mortality rate of 50 percent.<sup>12</sup> There is debate as to the appropriate management of perianal infections in this population, however, the presence of a necrotizing soft tissue infection is an indication for surgical debridement with rare exception.<sup>7</sup> Problems specific to this population include unique microbiology, lack of typical clinical findings, and profound immunosuppression.

There is an increased incidence of pseudomonal infections within the neutropenic population.<sup>1,7,11,13,14</sup> The theory behind the pathogenesis is an abnormal carriage of the pathogenic bacteria by cancer patients, with introduction into systemic circulation via translocation through damaged mucosa.<sup>14</sup> The presence of *Pseudomonas* bacteremia is often a preceding clinical clue prior to development of cutaneous infections.<sup>13-14</sup> A pathognomonic feature of this disease process is ecythma gangrenosum, often heralding further tissue destruction as a red inflamed lesion with a central black necrotic spot.<sup>14</sup> While our patient did not manifest this finding, this clinical feature is important to recognize within the neutropenic population, as the typical clinical presentation of these infections are often blunted to non-specific symptoms including localized pain and fever, followed by rapid progression and tissue destruction.<sup>1-2,4,7-9</sup> The role of neutropenia in the course of infections remains debated. While some studies have stated its importance regarding prognosis and operative decisions, the impact of neutropenia in necrotizing soft tissue infections is less clearly defined.<sup>4</sup> While our patient's immunosuppression undoubtedly was intimately related to her subsequent infection, there remains a paucity of literature to support the prognostic role of neutropenia post-operatively.



Another unique facet of this case is the subsequent creative reconstructive therapy that she will require. As the patient is young with a potentially curable malignancy, it is not unreasonable that she desires colostomy reversal in the future. Again, a lack of literature exists focusing on reconstruction following extensive debridement of the anal sphincters. Khalil et al described a single case of a young patient undergoing successful posterior sagittal anorectoplasty following wide debridement with partial sphincter involvement.<sup>14</sup> Additional considerations may include pelvic floor physical therapy, biofeedback, injectable bulking agents, sacral nerve stimulation, gracilis flaps, and other forms of neo-sphincters.

This case is noteworthy for several reasons. First, the vast majority of the literature regarding perianal sepsis amongst neutropenic patients focuses on those with leukemia, with few cases of Hodgkin lymphoma described. Next, the progression to a necrotizing infection, especially one involving the anal sphincters, is uncommon despite the relative frequency of perianal complications in this population. Lastly, this complication occurred in a pediatric patient with a potentially long life ahead of her. Not only is this important for the potential reconstructive options that will be addressed in a multidisciplinary fashion, but the psychological impact that such a traumatic experience can have on a young patient cannot be overlooked. Early recognition and debridement are key factors to minimizing the significant sequelae resultant from this disease process.

## Conclusion

Neutropenia associated with hematologic malignancy is a known risk factor for development of perianal sepsis. We present a case of a pseudomonal perianal necrotizing soft-tissue infection in an eleven-year-old female undergoing chemotherapy for Hodgkin lymphoma. This case highlights the need for high clinical suspicion and an early multidisciplinary approach in this population.

## Lessons Learned

Perianal complaints present not uncommonly in the neutropenic hematologic patient. Rarely, these can progress to necrotizing infections that can result in significant morbidity and mortality. High clinical suspicion and a multidisciplinary approach to these patients are critical.

## References

1. Grewal H, Guillem JG, Quan SHQ, Enker WE, Cohen AM. Anorectal disease in neutropenic leukemic patients: Operative vs. nonoperative management. *Dis Colon Rectum*. 1994;37(11):1095-1099. doi:10.1007/BF02049810
2. Baker B, Al-Salman M, Daoud F. Management of acute perianal sepsis in neutropenic patients with hematological malignancy. *Tech Coloproctol*. 2014;18(4):327-333. doi:10.1007/s10151-013-1082-z
3. Holzheimer RG, Mannick JA, eds. *Surgical Treatment: Evidence-Based and Problem-Oriented*. Munich: Zuckschwerdt; 2001. <http://www.ncbi.nlm.nih.gov/books/NBK6880/>
4. Büyükaşık Y, Özcebe OI, Saynalp N, et al. Perianal infections in patients with leukemia: Importance of the course of neutrophil count. *Dis Colon Rectum*. 1998;41(1):81-85. doi:10.1007/BF02236900
5. Runau F, Lane T, McMahan G, et al. Peri-Anal Sepsis in the Neutropenic Patient - Strategy Revisited. *J Surg Transplant Sci*. 2016;4(6):1044.
6. Sullivan P, Moreno C. A Multidisciplinary Approach to Perianal and Intraabdominal Infections in the Neutropenic Cancer Patient. *Oncology*. 2015;29(8).
7. Angel C, Patrick CC, Lobe T, Rao B, Pui C-H. Management of anorectal/perineal infections caused by *Pseudomonas aeruginosa* in children with malignant diseases. *J Pediatr Surg*. 1991;26(4):487-493. doi:10.1016/0022-3468(91)91001-F
8. Bublick MP, Hitchcock CR. Necrotizing anorectal and perineal infections. *Surgery*. 1979;86(4):655-662.
9. Glenn J, Cotton D, Wesley R, Pizzo P. Anorectal Infections in Patients with Malignant Diseases. *Clin Infect Dis*. 1988;10(1):42-52. doi:10.1093/clinids/10.1.42
10. Abercrombie JE, George BD. Perianal abscess in children. *Ann R Coll Surg Engl*. 1992;74(6):385-386.
11. Givler RL. Necrotizing anorectal lesions associated with *Pseudomonas* infection in leukemia: *Dis Colon Rectum*. 1969;12(6):438-440. doi:10.1007/BF02617729
12. Badgwell BD, Chang GJ, Rodriguez-Bigas MA, et al. Management and Outcomes of Anorectal Infection in the Cancer Patient. *Ann Surg Oncol*. 2009;16(10):2752-2758. doi:10.1245/s10434-009-0626-y
13. Pini Prato A, Castagnola E, Micalizzi C, et al. Early diverting colostomy for perianal sepsis in children with acute leukemia. *J Pediatr Surg*. 2012;47(10):e23-e27. doi:10.1016/j.jpedsurg.2012.05.034
14. Khalil BA, Baillie CT, Kenny SE, et al. Surgical strategies in the management of ecthyma gangrenosum in paediatric oncology patients. *Pediatr Surg Int*. 2008;24(7):793-797. doi:10.1007/s00383-008-2159-z