Unusual association of diseases/symptoms

Graft-versus-host disease

Joel A Newman,¹ Sophie Candfield,¹ David Howlett,² Paula McKenzie,² Satyajit Sahu¹¹ Department of Haematology, Eastbourne District General Hospital, Eastbourne, UK² Department of Radiology, Eastbourne District General Hospital, Eastbourne, UK

Correspondence to Joel A Newman, joel.newman@nhs.net

Summary

This article presents the case of a 26-year-old man who was diagnosed with pneumatosis cystoides intestinalis, a rare consequence of colonic graft-versus-host disease, after CT imaging revealed free air tracking within the bowel wall and abdomen. His presentation with hiccups, abdominal and neck distension, along with diarrhoea was also unusual, and this case highlights the need for a high index of suspicion when treating post stem cell transplant patients.

BACKGROUND

An increasing number of patients with both haematological malignancies and other life-threatening conditions are undergoing allogeneic stem cell transplantation. The side effects and sequelae experienced as a result of these procedures are not well known outside haematology circles, yet may present to any acute hospital. This case highlights the need for heightened clinical awareness when dealing with such patients, and the need for multidisciplinary input when caring for them.

CASE PRESENTATION

A 26-year-old man presented with a short history of neck swelling, hiccups and abdominal distension. He had ongoing watery bowel motions after being diagnosed 3 weeks earlier with acute graft-versus-host disease (GVHD) of the colon following an uneventful colonoscopic biopsy. In his medical history, he had undergone a matched unrelated allogeneic stem cell transplant for acute myeloid leukaemia 4 months previously. Medications included immunosuppression with cyclosporine A and prednisolone.

Figure 1 Axial CT imaging of the lower abdomen demonstrates changes with pneumatosis cystoides intestinalis, with air seen in the caecal wall (arrows). Some free intraperitoneal air is also present (arrowheads).

Figure 2 Axial CT imaging of the root of the neck shows extensive surgical emphysema with air tracking between muscle planes anteriorly and posteriorly.

On examination, he appeared well, with no fevers or haemodynamic compromise. Cardiorespiratory examination was unremarkable, and his abdomen was mildly distended, but soft and non-tender with no guarding or rebound. Bowel sounds were present and normal. Rectal examination revealed an empty rectum with no blood present. Routine blood tests had not significantly changed from results obtained 2 weeks previously. Plain abdominal radiographs were reported as unremarkable with no pathological dilatation of the bowel or free air seen. In view of his medical history, and persisting symptoms, axial CT imaging of the neck, chest, abdomen and pelvis was obtained for further investigation.

INVESTIGATIONS

CT imaging performed during the assessment of these symptoms demonstrated pneumatosis cystoides intestinalis (PCI) with tracking of gas through fascial planes, causing a pneumoperitoneum, pneumomediastinum and subcutaneous emphysema (see figures 1 and 2), a rare complication of the patient's biopsy-confirmed colonic GVHD.

TREATMENT

In our patient, methylprednisolone, cyclosporine A, tacrolimus, and later rituximab were initiated for treatment of his GVHD, along with supportive oxygen therapy, resulting in a complete recovery.

OUTCOME AND FOLLOW-UP

Complete resolution of the PCI was seen with ongoing treatment of the GVHD.

DISCUSSION

PCI is a rare condition where gas is found within mucosa and submucosa of the small or large intestine. Its patho-

physiology is unclear, and although it may be asymptomatic and primary, it generally occurs secondary to localised or systemic infective and inflammatory conditions, mechanical trauma, bowel ischaemia, chronic obstructive pulmonary disease or drugs. PCI is an important differential diagnosis for the cause of pneumoperitoneum, where the suspected pathology is rupture of an intestinal bleb which results in retroperitoneal free air tracking through fascial planes.

PCI often requires no specific treatment, but other treatment options include high dose antimicrobial therapy and aggressive oxygen therapy; surgical intervention is reserved for those patients with evidence of perforation, peritonitis or abdominal sepsis.^{1 2} PCI has rarely been associated with GVHD in the literature,³ and we believe these are the first published CT images of GVHD-associated PCI in the adult population.

Learning points

- Illnesses in patients after bone marrow transplantation require thorough investigation and consideration of a wide differential diagnosis.
- Although most haematological patients are admitted directly to specialist units, an awareness of the conditions affecting bone marrow transplant patients – such as graft-versus-host disease – is important for all frontline medical and surgical doctors.
- Multidisciplinary care is required for these patients, with radiologists knowledgeable about and experienced in dealing with the complications these patients face.
- Pneumatosis cystoides intestinalis is a differential diagnosis for the cause of pneumoperitoneum.

Competing interests None.

Patient consent Not obtained.

REFERENCES

1. **Zplke C**, Ulbrich S, Graeb C, *et al*. Acute pneumatosis cystoides intestinalis following allogeneic transplantation – the surgeon's dilemma. *Bone Marrow Transplant* 2002;**29**:795–8.
2. **Braumann C**, Menenakos C, Jacobi CA. Pneumatosis intestinalis—a pitfall for surgeons? *Scand J Surg* 2005;**94**:47–50.
3. **Day DL**, Ramsay NK, Letourneau JG. Pneumatosis intestinalis after bone marrow transplantation. *AJR Am J Roentgenol* 1988;**151**:85–7.

This pdf has been created automatically from the final edited text and images.

Copyright 2010 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Newman JA, Candfield S, Howlett D, McKenzie P, Sahu S. Graft-versus-host disease. *BMJ Case Reports* 2010;10.1136/bcr.05.2010.3033, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow