Evaluation of Abdominal Pain in the AIDS Patient

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Acquired immune deficiency syndrome (AIDS) is a recently recognized entity characterized by a deficiency in cell mediated immune response. The syndrome is manifested by the development of otherwise rare malignant neoplasms and severe life-threatening opportunistic infections. Case histories of five AIDS patients evaluated for abdominal pain are presented to demonstrate the unusual spectrum of intra-abdominal pathology that may be encountered in the AIDS patient. As the number of patients with AIDS continues to escalate, surgical evaluation and intervention will be required more frequently. An understanding of this syndrome and its complications is mandatory for the surgeon to adequately evaluate AIDS patients with abdominal pain.

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SINCE 1979, a new disease entity, the acquired immune deficiency syndrome (AIDS) has appeared in previously healthy individuals. This syndrome, characterized by a deficiency in cell mediated immune response, is manifested by the development of unusual malignant neoplasms and life-threatening opportunistic infections.[1-3]

To date, over 3000 cases have been documented and they continue to be reported to the Centers for Disease Control (CDC) at a rate of two to three new cases per day. Over 75% of the patients are male homosexuals. The remainder include primarily drug abusers, recipients of blood products, or Haitians. There is no effective therapy for the underlying immune defect at this time. Most patients die within two to four years of initial presentation, usually due to overwhelming opportunistic infections. A smaller proportion of these patients die due to progressive neoplastic processes.[4-6]

Severe abdominal pain in the AIDS patient is a diagnostic problem that increasingly elicits surgical consultation. However, the approach to this problem has received surprisingly little attention in the surgical literature. Five case histories of AIDS patients referred for surgical evaluation of abdominal pain are presented in the following report to demonstrate the unusual spectrum of pathologic processes that may be associated with abdominal pain in AIDS. Aspects of the AIDS syndrome that are particularly relevant for the surgeon involved in the evaluation of a patient with AIDS are emphasized.

Surgical evaluation and intervention will be required more frequently as the number of cases of AIDS continues to escalate. A thorough knowledge of the unusual infections and malignant processes that may be present in the AIDS patient is mandatory for the surgeon involved in the management of a patient with established or suspected AIDS.

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Materials and Methods

The medical records of five AIDS patients referred for surgical evaluation of severe abdominal pain were reviewed. All autopsies were reviewed by one of the authors. All patients met CDC criteria (Table 1) at the time of initial diagnosis of AIDS, which was 6 to 11 months prior to presentation with abdominal pain. Immunologic profiles in all five patients performed while they were ambulatory showed immunologic defects consistent with findings previously published.[1,2,6] Briefly, these defects included depressed blastogenesis to mitogens and specific antigens, lymphopenia with a selective decrease in T cells, and a marked depletion of the helper T lymphocytes as opposed to the suppressor T lymphocytes (OKT4/OKT8 ratio < 1.0).

Case Reports

Case 1. A 37-year-old white male homosexual had been healthy until he developed *Pneumocystis carinii* pneumonia. A thorough evaluation revealed no other infection or neoplasm and he responded to treatment with intravenous trimethoprim–sulfamethoxazole. He was asymptomatic for the following 11 months until he was admitted to the surgical service with an acute onset of sharp right upper quadrant abdominal pain radiating to the back and to the left upper quadrant. The pain was accompanied by frequent episodes of watery diarrhea, nausea, and vomiting.

His past history was significant for an episode of intestinal amebiasis 2 years previously, which resolved with appropriate antimicrobial therapy. Three months prior to admission he had a self-limited episode of bright red bleeding per rectum. Evaluation at that time included a barium

enema and colonoscopy with multiple biopsies revealing mild nonspecific mucosal inflammation.

On admission to the surgical service, the patient's temperature was 103 F orally. There were no cutaneous lesions suggestive of Kaposi's sarcoma. The abdomen was nondistended with diminished bowel sounds present. Severe tenderness with marked rebound and involuntary guarding was present in both lower quadrants. No masses were noted. Rectal exam was unremarkable and the stool was guaiac negative. The leukocyte count was 3600/mm^3 and other routine laboratory studies were within normal limits. A chest x-ray was unremarkable. Abdominal films showed a nonspecific gas pattern and there was no evidence of free air in the abdomen.

An emergency laparotomy was undertaken with a preoperative diagnosis of a perforated viscus. A midileal perforation measuring 3 mm in diameter was identified on the antimesenteric border of the ileum, surrounded by inflamed necrotic tissue. This was resected and pathologic examination revealed a perforated, poorly differentiated lymphoma, with four of four mesenteric lymph nodes involved with tumor. Postoperative staging evaluation revealed no other evidence of lymphoma. He was treated with six cycles of adriamycin, cytoxan, vincristine, and prednisone at four weekly intervals. He remained disease-free until 9 months after surgery when he presented with disseminated *Mycobacterium avium-intracellulare,* which was cultured from urine, sputum, and bone marrow biopsy specimens. He died shortly after this diagnosis was established and an autopsy was not performed.

Case 2. A 48-year-old black male bisexual developed candida esophagitis and cutaneous Kaposi's sarcoma. During treatment for candida esophagitis, he was noted to have *M. avium-intracellulare* in his sputum. His chest x-ray was normal at that time and he had no evidence of active disease so antimycobacterial therapy was not initiated. Eight months later, he presented with a 2-week history of fever and diffuse abdominal pain. He denied nausea, vomiting, anorexia, rectal bleeding, or a change in bowel habits. His past medical history was unremarkable.

On initial presentation his temperature was 101.4 F orally. Abdominal examination revealed mild diffuse tenderness with no localization, guarding, or masses. Abdominal computerized tomography, oral cholecystogram, barium enema, and upper gastrointestinal series were remarkable only for para-aortic lymphadenopathy and biliary sludge. A chest x-ray showed a left lower lobe infiltrate. Sigmoidoscopy showed nonspecific proctitis. Complete blood count and chemistry profile were remarkable only for a leukocyte count of 3000/mm^3. Cultures of blood, urine, and stool were negative for pathogenic bacteria. Sputum cultures grew only *Staphylococcus aureus.* The patient was treated with clindamycin, penicillin, vancomycin, gentamicin, and ketoconazole for presumed pulmonary sepsis for 3 weeks without resolution of his symptoms. He was then transferred to the National Institutes of Health.

After transfer, the patient reported an exacerbation of his abdominal pain with localization to the right upper quadrant. His temperature was 102 F orally. Mild abdominal distention and hypoactive bowel sounds were noted. Generalized abdominal tenderness was present, most severe in the right upper quadrant. Right costovertebral angle tenderness was present. Rectal examination revealed minimal external anal tenderness and the stool was guaiac negative. Leukocyte count was 2000/mm^3, lactate dehydrogenase (LDH) was 491 U/L (normal 133–248 U/L), and serum glutamic acid oxaloacetate transaminase (SGOT) was 39 U/L (normal 8–31 U/L). A chest x-ray, abdominal flat and upright films, and intravenous pyelogram were normal. With a preoperative diagnosis of cholecystitis, an emergency laparotomy was performed, which revealed only hepatosplenomegaly. There was no evidence of biliary or other intra-abdominal pathology. A liver biopsy specimen showed moderate inflammation with intranuclear inclusion cells characteristic of cytomegalovirus. His abdominal pain appeared to be due to cytomegalovirus hepatitis.

Five days after surgery, the patient developed diffuse pulmonary infiltrates. An open lung biopsy showed *P. carinii* and cytomegalovirus inclusion cells. Despite the initiation of pentamidine and trimethoprim-sulfamethoxazole therapy, the patient died due to progressive hypoxemia.

TABLE 1. *Acquired Immune Deficiency Syndrome: Centers for Disease Control (CDC) Definition**

Based on clinical, not laboratory evidence

A. Patient with no known cause for immunosuppression, and

B. Disease predictive of a defect in cellular immune function

1. Pneumonia, meningitis or encephalitis due to:

Aspergillus	*Nocardia*
Cytomegalovirus	*Toxoplasma*
Strongyloids	*Cryptococcus*
Atypical mycobacteria	*Pneumocystis*
Candida	*Zygomycosis*

2. Esophagitis due to *Candida,* Herpes simplex, cytomegalovirus
3. Progressive multifocal leukoencephalopathy
4. Chronic enterocolitis due to cryptosporidiosis
5. Extensive Herpes simplex > 5 weeks
6. Kaposi's sarcoma
7. NonHodgkin's central nervous system lymphoma

* CDC criteria providing a strict, working definition of AIDS continue to be amended frequently as information accumulates. These criteria are necessarily restrictive to prevent possible inclusion of inappropriate cases in CDC statistics. It is widely recognized that many patients with AIDS may not fulfill present CDC criteria, though clinical, epidemiologic, and immunologic data can be compelling that they, in fact, do have AIDS.

Postmortem examination revealed extensive cytomegalovirus infection in the duodenum, pancreas, spleen, liver, kidneys, and adrenal glands. The spleen weighed 240 grams, and acid fast bacilli were identified on tissue touch preparations; *M. avium-intracellulare* was cultured from the spleen. No other foci of infection with *M. avium-intracellulare* were identified. There was no evidence of visceral involvement with Kaposi's sarcoma.

Case 3. A 35-year-old white male homosexual had developed *P. carinii* pneumonia and recurrent candida esophagitis 6 months prior to presentation with disseminated mycobacterial infection. *Mycobacterium avium-intracellulare* grew from sputum, lung biopsy, urine, and bone marrow specimens. Therapy was initiated with clofazimine, rifampin, and isoniazid. During this hospitalization he developed a sudden onset of severe generalized abdominal pain. This was not associated with vomiting, diarrhea, or nausea. His past medical history was remarkable only for two episodes of hepatitis in the past 7 years, and for an appendectomy during childhood.

He was febrile to 102.8 F orally. On physical examination, his abdomen was severely distended with hypoactive bowel sounds, diffuse tenderness to palpation, and involuntary guarding. The leukocyte count, which had ranged from 2000 to 4000/mm^3 since admission, was 7600/mm^3 and other routine laboratory studies were unchanged. A chest x-ray was unremarkable. Flat and upright abdominal films revealed a gas pattern suggestive of a mild ileus but were otherwise unremarkable. An emergency exploratory laparotomy was undertaken, revealing a massively enlarged spleen and a 12 × 7.5 cm mesenteric mass at the base of the small bowel mesentery, initially thought to be neoplastic. The mass was resected, and the liver and mesenteric lymph nodes were biopsied. Acid fast bacilli were identified by microscopic examination in all biopsy specimens; there was no evidence of neoplasm. Subsequent culture of biopsies of the mesenteric mass, the liver, and multiple enlarged mesenteric lymph nodes all grew *M. avium-intracellulare.* After surgery, the patient initially had an uneventful recovery. He continued to be treated for disseminated mycobacterial disease and had no recurrence of abdominal pain. One

TABLE 2. *Causes of Abdominal Pain in Acquired Immune Deficiency Syndrome*

Etiology	Associated Abdominal Manifestations/Complications	Diagnostic Studies*
Candida albicans	Esophagitis, enterocolitis, gastrointestinal bleeding	Endoscopy, biopsy, and culture
Cryptococcus neoformans	Intra-abdominal inflammatory mass, peritoneal seeding	Biopsy and culture
Histoplasma capsulatum	Hepatosplenomegaly, retroperitoneal lymphadenopathy	Biopsy and culture
Mycobacterium avium-intracellulare	Hepatosplenomegaly, intra-abdominal inflammatory mass, diarrhea, enterocolitis, peritoneal seeding, retroperitoneal lymphadenopathy	CAT scan, biopsy, and culture
Cytomegalovirus	Gastrointestinal bleeding, diarrhea, enterocolitis, hepatitis, esophagitis	Biopsy
"Gay Bowel Syndrome"†	Gastrointestinal bleeding, diarrhea, proctitis, enterocolitis, intestinal perforation	Endoscopy, biopsy and culture, and ova/parasite examination
Kaposi's sarcoma	Gastrointestinal bleeding, retroperitoneal lymphadenopathy, intestinal obstruction	CAT scan, endoscopy, and biopsy
NonHodgkins' lymphoma	Gastrointestinal bleeding, intestinal perforation, retroperitoneal lymphadenopathy	CAT scan and Biopsy

* Biopsy specimens may be obtained by upper or lower gastrointestinal endoscopy, laparoscopy, laparotomy, or by CAT-directed needle biopsy depending on suspected organ involvement and patient's condition.

† Gay Bowel Syndrome refers to proctocolitis due to trauma or to a wide range of enteric organisms including Herpes simplex, Salmonella, Shigella, Campylobacter, Entamoeba histolytica, Cryptosporidium, Giardia lamblia, Chlamydia, Neisseria gonorrhoeae, Treponema pallidum.

month after his abdominal exploration, he developed *P. carinii* and cytomegalovirus pneumonia and died due to respiratory failure.

Postmortem examination revealed *M. avium-intracellulare* in the spleen, kidneys, liver, lymph nodes, and lungs. Cytomegalovirus inclusion cells consistent with active infection were demonstrated in the spleen, esophagus, stomach, duodenum, liver, pancreas, adrenal glands, kidneys, lymph nodes, and lungs. The small bowel mesentery appeared normal. There was no evidence of cutaneous or visceral Kaposi's sarcoma.

A malignant lymphoma of the diffuse, large cell type was identified in the liver. An extensive search failed to reveal lymphomatous involvement of any other organs.

Case 4. A 35-year-old white male homosexual with a 7-month history of candida esophagitis, cytomegalovirus viremia, and Kaposi's sarcoma involving lymph nodes, skin, and mucous membranes of oropharynx presented with ataxia, hemiparesis, and a global dementia. Brain biopsy of a parietal lobe mass demonstrated foci of demyelinization but no etiologic agent was identified. Ten days after the brain biopsy, the patient developed mild abdominal tenderness which was continuous and associated with bloody diarrhea. He remained afebrile, but became anorectic and developed hepatomegaly. A liver spleen scan demonstrated an enlarged liver with multiple areas of decreased uptake. An abdominal ultrasound showed hepatomegaly without focal defects. Flat and upright abdominal films revealed a gas pattern consistent with colonic ileus. A chest x-ray was unremarkable. Microbiologic studies of blood, sputum, urine, and stool showed only cytomegalovirus viremia. The patient's abdominal tenderness resolved spontaneously 10 days after the initial onset, but recurred 1 week later. At this time, it was associated with nausea and frequent vomiting of bilious material. Multiple new cutaneous lesions of Kaposi's sarcoma were evident on the patient's face, neck, and extremities. An upper gastrointestinal series was performed, which was essentially normal. Chemotherapy for progressive Kaposi's sarcoma was started (vinblastine, adriamycin, bleomycin, vincristine, actinomycin D, and dimethyl-triazeno-imidazole-carboxamide). Oral lesions of Herpes simplex developed and were treated with intravenous Acyclovir.® In the following week, the patient developed progressive abdominal distention associated with continued bloody diarrhea, anorexia, and nausea. His temperature peaked at 101.4 F orally and he subsequently defervesced spontaneously. A surgical consultation was obtained. At that time, he appeared cachectic but in no acute distress. The abdomen was distended and mildly tender to palpation with voluntary guarding, but there was no rebound tenderness. Hypoactive bowel sounds were present. Hepatomegaly was appreciated but there were no splenomegaly or other abdominal masses palpable. The leukocyte count was 2900/mm^3, LDH was 393 U/L; SGOT was 37 U/L, and serum albumin was 2.8 gm/dl (normal 3.8–4.9 gm/dl). A chest x-ray was unremarkable. Abdominal flat and upright films revealed a nonspecific gas pattern without evidence of intestinal obstruction. Sigmoidoscopy to 25 cm showed multiple lesions consistent with Kaposi's sarcoma in the sigmoid colon and rectum. After an essentially negative microbiologic evaluation, his abdominal pain and bleeding were thought to be manifestations of extensive lower gastrointestinal involvement with Kaposi's sarcoma. Total parenteral nutrition was initiated and chemotherapy for progressive Kaposi's sarcoma was continued. Persistent lower gastrointestinal bleeding required multiple blood transfusions. There was only marginal improvement in the patient's abdominal complaints during total parenteral nutrition and complete bowel rest. The cutaneous Kaposi's sarcoma lesions progressed on chemotherapy and the guaiac positive diarrhea persisted with repeatedly nondiagnostic microbiologic evaluations. Two weeks after initial surgical consultation the patient developed *P. carinii* pneumonia, diagnosed by open lung biopsy. Despite therapy with pentamidine, he died due to respiratory failure.

Postmortem examination revealed extensive intra-abdominal Kaposi's sarcoma with numerous ulcerated and nonulcerated lesions identified throughout the submucosa of the esophagus, duodenum, small intestine, appendix, and colon. The liver weighed 1500 grams and had multiple lesions of Kaposi's sarcoma measuring up to 1.5 cm in diameter scattered throughout the hepatic parenchyma. Kaposi's sarcoma was also identified in the gallbladder, biliary ducts, pancreas, and the undersurface of the diaphragm.

Case 5. A 42-year-old white male homosexual with a 6-month history of Kaposi's sarcoma and oral candidiasis complained of intermittent right lower quadrant and right groin pain during a routine clinic visit. At the time he was receiving therapy with lymphoblastoid alpha interferon (Burroughs-Wellcome, Research Triangle, NC) for progressive cutaneous Kaposi's sarcoma. The past medical history was remarkable only for bilateral inguinal hernia repairs 7 years previously. The pain was associated with diarrhea but no nausea or vomiting. An abdominal ultrasound and stool cultures were unremarkable. Abdominal computerized tomography was within normal limits. One week later the patient again noted severe right groin and lower abdominal pain associated with a 2 × 2 cm right groin mass. He was afebrile. On physical examination, multiple cutaneous lesions of Kaposi's sarcoma on his face, neck, and

shoulders were identified. An irreducible right inguinal hernia was present, associated with moderate right lower quadrant tenderness and voluntary guarding.

The patient was taken to the operating room for reduction of an incarcerated hernia. Inspection of the contents of the hernia sac revealed inflamed omental tissue, firmly adherent to the right spermatic cord. A laparotomy was then performed, revealing a firm, thickened, nodular omentum and multiple peritoneal nodules studded throughout the abdominal cavity. This was initially thought to represent a neoplastic process, but microscopic examination of omental and peritoneal nodule biopsies revealed the presence of encapsulated yeast with no malignant cells identified.

A right herniorraphy was performed and *Cryptococcus neoformans* subsequently grew out of all biopsied tissues. After surgery, the patient was treated with flucytosine and intravenous amphotericin B for disseminated *C. neoformans*. Four weeks after laparotomy he developed *P. carinii* pneumonia. Three months later, he developed gastrointestinal bleeding from gastric Kaposi's sarcoma lesions. His right lower quadrant and right groin pain have not recurred after surgery.

Discussion

Evaluation of the AIDS patient with abdominal pain presents a unique diagnostic challenge to the surgeon. Correlation of the clinical courses of patients presented in this report with the findings at laparotomy and autopsy highlights the unusual spectrum of disease processes that must be included in the differential diagnosis of abdominal pain in these patients (Table 2). These case histories also demonstrate the difficulties encountered in fully evaluating and treating abdominal pathologic processes in AIDS patients.

There is currently no unequivocal serologic or immunologic marker to identify AIDS patients. The Centers for Disease Control define a case of AIDS as a disease at least moderately predictive of a defect in cell mediated immunity occurring in a person with no known cause for diminished resistance to the disease (Table 2).[7] Patients with AIDS are particularly susceptible to the development of an unusual spectrum of infectious processes (Table 3) which may be associated with abdominal pain.

Infection with cytomegalovirus can be documented in virtually every patient with the diagnosis of AIDS. However, the etiologic, pathophysiologic, or prognostic significance of asymptomatic cytomegalovirus infections in this population is an unresolved issue which currently remains among the most widely debated aspects of this syndrome.[8-10] Cytomegalovirus can induce transient abnormalities in cellular immune function in otherwise healthy persons, associated with a mononucleosis-like illness. It can also cause disseminated disease in many immunosuppressed patient populations.[11-13] Typically, an AIDS patient with cytomegalovirus disease presents with fever, malaise, and lymphadenopathy. Hepatitis, pancreatitis, gastrointestinal ulceration, bleeding, and perforation are among the manifestations of intra-abdominal cytomegalovirus infection that have been reported in immunosuppressed patients.[14-19] However, the role of disseminated cytomegalovirus infection in the development of abdominal pain in the AIDS patient has not been established.

TABLE 3. *Infectious Complications of AIDS*

Frequent	Rare
Pneumocystis carinii	*Listeria*
Mycobacterium avium-intracellulare	*Nocardia*
Herpes simplex	
Candida	*Mucor*
Cryptococcus neoformans	*Aspergillus*
Toxoplasma gondii	
Cytomegalovirus	
Cryptosporidium	
Mycobacterium tuberculosis	

Two of the five patients in this series presenting with abdominal pain had disseminated cytomegalovirus infections documented at autopsy. Because both of these patients had evidence of additional intra-abdominal pathologic processes, the role of the disseminated cytomegalovirus infection alone in the production of abdominal pain cannot be specifically defined. However, hepatic cytomegalovirus involvement with characteristic inflammatory changes was present in both cases and has been associated with diffuse abdominal pain in other immunosuppressed patients.

The diagnosis of active cytomegalovirus infection in the abdomen requires tissue biopsy with the demonstration of inclusion cells or isolation of virus from the affected organ in the presence of a compatible histologic response. High cytomegalovirus titers alone are not diagnostic or predictive of the presence of intra-abdominal cytomegalovirus infection. Although patients with active cytomegalovirus infections have been treated with Acylovir, this has not substantially affected the course of the disease and there is currently no adequate treatment available for disseminated cytomegalovirus infection.[7]

AIDS patients are susceptible to the development of *M. avium-intracellulare* infections but apparently not to other atypical mycobacterial diseases.[20] Prior to the emergence of AIDS, *M. avium-intracellulare* had been described primarily in middle-aged men with obstructive pulmonary disease, causing a chronic, localized cavitary pulmonary disease. Disseminated *M. avium-intracellulare* has rarely been described in other immunocompromised patients.[21-23] An increased frequency of disseminated *M. avium-intracellulare* infections in the AIDS population has been reported by several groups studying this syndrome.[20,24,25]

Mycobacterium avium-intracellulare can clearly be implicated in the development of acute abdominal pain

resulting in emergency laparotomy in one of the cases in this report. In Case 3, laparotomy was required to diagnose extensive intra-abdominal *M. avium-intracellulare* infection and to exclude the presence of any other acute process requiring surgical intervention.

There were two other patients in this series with *M. avium-intracellulare.* The patient in Case 1 died with disseminated *M. avium-intracellulare* while apparently in remission with nonHodgkin's lymphoma. An autopsy was not performed so the extent of possible asymptomatic intra-abdominal involvement with *M. avium-intracellulare* is unknown. The patient in Case 2 had splenic infiltration with *M. avium-intracellulare* at autopsy. Although splenomegaly had been noted at laparotomy, no splenic tissue was obtained for pathologic or microbiologic examination at that time. This visceral intra-abdominal disease was not suspected clinically so the patient was never treated with antimycobacterial therapy. It is interesting to note that, although sputum cultures several months earlier had grown *M. avium-intracellulare,* the spleen was the only organ involved with this infection at autopsy. The significance of this infection in the development of diffuse abdominal pain is unknown. The AIDS patient in this case never had localized left upper quadrant symptoms during his hospitalization.

The diagnosis of *M. avium-intracellulare* depends on demonstration of acid-fast organisms in appropriately stained biopsy specimens, confirmed by culture of *M. avium-intracellulare* from biopsy specimens. The pathologic examination of tissues from AIDS patients with extensive *M. avium-intracellulare* infections is frequently striking for the relative paucity of tissue response despite the presence of innumerable acid fast bacilli.[7,25] Acid-fast stains should be performed routinely on any tissue specimen obtained from these patients even in the absence of characteristic caseation and granuloma formation. This is particularly important when mycobacterial infection is not suspected clinically. The treatment of disseminated *M. avium-intracellulare* in AIDS patients with conventional antimycobacterial regimens has been uniformly unsuccessful so far and the patient's prognosis with disseminated disease is presently very poor.[7,20] Two investigational drugs, Clofazamine (National Hansen's Disease Center, Carville, LA) and Ansamycin (Centers for Disease Control, Atlanta, GA), which have had good *in vitro* activity against *M. avium-intracellulare,* are currently being used in the treatment of AIDS patients with disseminated *M. avium-intracellulare* infection.

The patient in Case 5 had disseminated intra-abdominal cryptococcosis, presenting as an incarcerated inguinal hernia. Cryptococcal infection is frequently seen in association with cell mediated immunodeficiency, although it can also occur in otherwise apparently healthy individuals. The presumed portal of entry for this airborne organism is the respiratory tract. Infection with this encapsulated yeast can present as a subacute or chronic illness. Subclinical infection may be relatively common in immunocompetent individuals, based on the results of extensive skin testing studies, but progressive disease seldom follows inhalation of the organism.[26] In the United States this infection has primarily been seen in patients with other serious underlying illnesses, including sarcoidosis, Hodgkin's disease, systemic lupus erythematosus, and in patients receiving corticosteroids following renal transplantation. Recent series have clearly documented that the most common foci of involvement are the meninges and the lungs. Although gastrointestinal involvement is unusual, the liver, spleen, and adrenal glands are apparently the organs most frequently involved when intra-abdominal involvement is present.[27-30] Cryptococcal peritonitis has also been reported.[31,32] Among infected AIDS patients, the organism can often be isolated from the cerebrospinal fluid, urine, sputum, and blood. The most common manifestations of cryptococcal infection in the AIDS patient include headache, fever, and disorientation, usually reflecting central nervous system involvement.[7] Although extensive intra-abdominal involvement has been documented in many patients with cryptococcal infection, the initial manifestation of intra-abdominal cryptococcus as an incarcerated inguinal hernia has not previously been reported.

The diagnosis of *C. neoformans* infection can be made by demonstration of the organisms' capsule on India ink examination of cerebrospinal fluid, peritoneal fluid, or sputum and confirmed by cultivation.[26] Current recommended treatment consists of more than 2 grams of intravenous amphotericin B, usually in combination with oral flucytosine. However, flucytosine may be poorly tolerated because of its myelosuppressive effects in AIDS patients who are chronically leukopenic.

Acquired immune deficiency syndrome patients are also more likely to develop unusual malignancies, particularly Kaposi's sarcoma and nonHodgkin's lymphoma, which the surgeon must consider in his differential diagnosis of abdominal pain (Table 2).

Until recently, Kaposi's sarcoma was a malignancy rarely seen in the United States,[33] although an increased incidence of the disease had been noted in renal transplant patients on immunosuppressive therapy.[34,35] The classical form of this disease was first described by Kaposi in 1872 as a localized, nodular skin tumor, ranging in color from blue to purple, on a lower extremity.[36] This neoplasm has generally been radiosensitive or amenable to chemotherapy with a long-term reported survival of 8 to 13 years.[37-39] Gastrointestinal tract involvement has been present in up to 50% of all cases studied, although it is

generally asymptomatic.[40–42] However, gastrointestinal Kaposi's sarcoma has been reported to cause bleeding, intestinal obstruction, and intussusception.[43–47]

The risk of development of Kaposi's sarcoma in homosexual males since the recognition of AIDS has been estimated by the Centers for Disease Control to be approximately 100 times greater than that for the general population.[48] The type of Kaposi's sarcoma that develops in the AIDS patient is frequently a far more virulent, aggressive form which may rapidly disseminate throughout the skin, mucous membranes, and gastrointestinal tract, and occasionally spread to the liver, spleen, and lungs.[46] The majority of AIDS patients with Kaposi's sarcoma also have readily visible skin lesions.[49,50]

The patient presented in Case 4 was found to have extensive gastrointestinal tract involvement with Kaposi's sarcoma at autopsy. This case demonstrates the rapidly progressive, frequently fatal outcome of this disease in AIDS patients. Radiographic studies may appear normal in patients with extensive gastrointestinal involvement and endoscopic evaluation with biopsy documentation appears to be a more sensitive method of diagnosis of gastrointestinal Kaposi's sarcoma.[42] Treatment modalities available for patients with disseminated Kaposi's sarcoma are limited. Occasionally, isolated lesions forming the focus for intussusception or intestinal obstruction have been surgically resected.[44,47] Cutaneous lesions have been treated successfully with chemotherapy or radiotherapy. Disseminated disease has been treated with chemotherapy, which can frequently be complicated in AIDS patients by severe opportunistic infections. A complete response rate in 15% of AIDS patients with disseminated Kaposi's sarcoma treated with doxorubicin, bleomycin, and vinblastine has been reported, though only short-term follow-up is available.[6]

An increased frequency of nonHodgkin's lymphomas has also been evident among AIDS patients, including central nervous system lymphomas (now part of CDC criteria for the diagnosis of AIDS, Table 2) and Burkitt's-like lymphoma.[51–53]

In Case 1, the patient's abdominal pain was caused by a perforated ileal lymphoma. In this case, the diagnosis of lymphoma could only be made at laparotomy, which was undertaken emergently because of the patient's acute presentation. In retrospect, this may have been the focus for the episode of lower gastrointestinal bleeding that the patient had 3 months prior to his presentation with abdominal pain. This predisposition to the development of unusual, potentially treatable malignancies such as lymphoma mandates a thorough evaluation including biopsy, if necessary, to establish a definitive diagnosis of any abdominal mass in an AIDS patient. Treatment for nonHodgkin's lymphoma will usually include combination chemotherapy. Specific recommendations would depend on histology and stage of disease at the time of diagnosis. Unfortunately, as this case demonstrates the long-term prognosis for the AIDS patient with lymphoma is poor even if the malignancy is treated. In Case 3 in this report, large cell histocytic lymphoma was found in the liver at autopsy. This was not suspected antemortem and demonstrates the unusual extent of intra-abdominal pathology that may remain clinically occult in these patients.

TABLE 4. *Precautions for Health-care Workers*

Staff Restrictions
No immunosuppressed workers
No pregnant workers (cytomegalovirus, hepatitis)
Contact Isolation (Gown and Glove)
Secretions
Excretions
Blood
Patients with poor hygiene
Respiratory Precautions
Active pneumonitis
Laboratory and Delivery Precautions
All specimens clearly labelled
Double bag specimens in impermeable sacs
Personnel gown and glove to protect against spatter
Work areas cleaned with Clorox

As these cases demonstrate, there are notable features of the AIDS patient that can contribute to difficulties encountered by the surgeon in diagnostic and therapeutic management decisions. The hospitalized AIDS patient will frequently present with multiple, disseminated infections and will have positive serology for cytomegalovirus and hepatitis A and B. The presence of severe concurrent pneumonia or other systemic illnesses often impedes the appropriate workup for abdominal complaints. As several of these cases illustrate, standard radiographic evaluations may frequently be unremarkable despite the presence of extensive intra-abdominal pathology.

For nonemergent evaluation of abdominal pain, a thorough workup of the patient's complaints will better define the role and nature of possible operative intervention. Careful consideration of the unusual malignant and infectious etiologies of abdominal pain that are more commonly encountered in this patient population will further direct appropriate diagnostic studies (Table 2). It must be emphasized that these studies should supplement but not replace an appropriate differential diagnostic approach that is applied to the evaluation of any patient with abdominal pain.

At laparotomy, undertaken either emergently or electively, adequate tissue specimens must be obtained by the surgeon for both microbiological and histological examination. Grossly, the malignant or infectious nature of a disseminated intra-abdominal process may be difficult

to assess. Immediate inspection of frozen section biopsies for histologic diagnosis and specially stained biopsy specimens for microbiologic studies is particularly helpful under these circumstances.

The infectivity of AIDS patients is an issue of particular concern to the surgeon. Since the vectors of transmission have not yet been clearly defined, it is impossible to give precautionary recommendations with absolute certainty. At the National Institutes of Health, all surgical procedures are performed with double gloving by the operating team and ancillary personnel. All pathology, microbiology, and laboratory specimens obtained from AIDS patients are handled as potentially highly infective material, and are clearly labelled as "AIDS PROTOCOL" specimens. Laboratory workers are encouraged to handle specimens with gown and glove precautions (Table 4). To date, no health-care worker has acquired AIDS from caring for an AIDS patient. Caution has to be exercised with regard to transmission of the AIDS agent, as well as infections such as cytomegalovirus, hepatitis A and B, and Herpes virus which so often accompany AIDS itself. The AIDS syndrome appears to be transmitted by sexual contact or by blood products, and rarely, if ever, by any other type of contact. Surgeons must be particularly careful to avoid penetrating needle or scalpel wounds from AIDS patients despite the fact that such accidental puncture has never been documented to transmit AIDS.[54-56]

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