CASE REPORT

A Case of Severe Hidradenitis Suppurativa Contributing to a Death and a Review of the Literature

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Abstract: Objective. Our aim was to report a fatal complication of hidradenitis suppurativa and a review of the literature. Materials and Methods. A case of severe hidradenitis suppurativa obscuring the diagnosis and precluding the treatment of a ruptured sigmoid diverticulum is presented. Results. We describe a 48-year-old woman who had a long history of untreated hidradenitis suppurativa with extensive vulvar involvement and poor nutritional status. The advanced state of her disease on initial presentation led directly to her death. Conclusion. Although it is a treatable disease, hidradenitis suppurativa can lead to systemic sequelae severe enough to contribute directly to death.

Key Words: folliculitis, hidradenitis suppurativa

Chronic hidradenitis suppurativa results from progressive inflammation and tissue damage associated with recurrent deep folliculitis in the axilla, periareolar, and perianal region [1]. Velpeau first described superficial inflammation of the skin appendages in 1839. In 1854, Verneuil recognized that these supplicative lesions originate in the sweat glands, but not until 1922 did Schiefferdeker associate this condition with the apocrine glands. In its severe form, the disease may result in chronic supplicative follicular lesions involving the adjacent skin appendages, forming sinuses and fistulae.

Early diagnosis and recognition of the disease may allow for effective local treatment. Failure of early diagnosis may allow incapacitation, responding only to radical surgery. Death from this benign disease has been reported [2, 3]. We present a severe case of a 48-year-old woman with untreated hidradenitis suppurativa of the perineum.

CASE REPORT

A 48-year-old G1P1 had increasing lower-extremity edema resulting in inability to walk and work at her job as a desk clerk. Her local physician suspected a malignancy and referred her to the Gynecologic Oncology Service at The University of Iowa Hospitals and Clinics.

On admission to the hospital, her review of systems showed fatigue, anorexia, a 30-pound weight loss over the last 6 months, difficulty in urinating, diarrhea for the prior 2 months, and a 20-year history of nodular
Hidradenitis suppurativa affects areas in which apocrine glands are located, including the axilla (61%), the groin (48%), the perineal (30–50%) and perirectal (22%) areas, and the breast (13%) [4]. Hidradenitis suppurativa occurs most often in the second to the fourth decade of life. Apocrine glands become active during puberty; thus, rarely does hidradenitis suppurativa occur during prepubescence. The incidence is approximately 1 in 300, although the exact number is un-

vulvar and axillary pustules that intermittently spontaneously drained. Twenty years prior to her presentation at our clinic, a dermatologist told her that she had a very rare, benign disease. Family history was negative for carcinoma.

On physical examination, she was a cachectic, frail, white woman appearing older than her stated age. Her weight was 43 kg, and her height was 158 cm. She was afebrile, with stable vital signs. Her abdomen was distended slightly, with active bowel sounds. The abdomen was tympanic to percussion without guarding or rebound. Her lower extremities had + pitting edema to the groin. The suprapubic and perineal areas were replaced by numerous sinus tracts and fistulae extending deep into the subcutaneous tissues. The pubic peritoneum was visible. The urethra was in the correct anatomical position, but locating the anus was difficult. Posteriorly, numerous draining sinus tracts extended toward the sacrum. The cervix was palpable, but the uterus and adnexa were not.

Laboratory values showed a white blood cell count of 25,000 mm$^3$, with 23,368/mm$^3$ neutrophils and 508/mm$^3$ bands. Her hemoglobin and hematocrit were 9.4 g/dL and 29%, respectively. Her platelet count was 577,000 mm$^3$. Serum sodium was 128 mEq/L, blood urea nitrogen was 14 mg/dL, and creatinine was 2.8 mg/dL. Total protein was 3.9 g/dL, with an albumin of 1.3 g/dL. Her calculated creatinine clearance was 18 mL/min. She had a fractional excretion of sodium of 0.30%.

Her chest radiogram showed free air under the diaphragm. An abdominal and pelvic computed tomography scan revealed hepatomegaly with fatty infiltration, edematous and thickened small bowel, and abdominal and pelvic free fluid. Renal ultrasonography showed no evidence of hydronephrosis.

Because we could visualize the pubic bone, we believed that the multiple fistula tracts accounted for the free air. Our differential diagnosis of hidradenitis suppurativa or Crohn’s disease was confirmed by the gastrointestinal service, which believed that the woman’s clinical picture was consistent with Crohn’s disease. Immediate surgical exploration was not offered because of her multisystem organ failure and generalized debilitated condition. She was transfused with two units of packed red blood cells and was started on antibiotics and total parenteral nutrition.

On hospital day 4, the patient became disoriented. Her abdomen was diffusely tender, and now she had guarding bilaterally in the lower quadrants. Her mental status declined progressively, and she became obtunded on hospital day 5. At that time, her temperature was 35.5°C, though she had been afebrile during her hospitalization. Laboratory findings included a serum ammonia of 109 μmol/L. A chest radiogram showed a new right lower-lobe infiltrate.

The patient was transferred to the intensive care unit for treatment of sepsis. Lumbar puncture was negative for white blood cells. Computed tomography scan of the head was negative. Although the patient had no seizure activity, an electroencephalogram was consistent with status epilepticus. On hospital day 6, she developed fixed and dilated pupils and Kussmaul’s respirations and later expired.

**PATHOLOGY**

A postmortem examination limited to the chest and abdomen showed sigmoid colon diverticulitis, a perforated sigmoid diverticulum, and peritonitis with abundant fibrinopurulent material covering the peritoneal walls and organs. Additionally, pneumonia of the right upper and right middle lung lobes was confirmed.

A 12 × 6 × 5-cm protuberant vulvar lesion with multiple sinus tracts covered the vulva and perineum and extended onto the lower abdominal wall, bilateral inner thighs, anus, and sacrococcygeal region (Fig. 1). The mass obscured the urethral meatus and anal orifice, though the vaginal introitus was readily identifiable. The lesion formed a polypoid tumorous mass covered by mottled skin with numerous fistula tracts and areas of scarring (Fig. 2). Fistula tracts communicating with the peritoneal cavity could not be established.

**DISCUSSION**

Hidradenitis suppurativa occurs most often in the second to the fourth decade of life. Apocrine glands become active during puberty; thus, rarely does hidradenitis suppurativa occur during prepubescence. The incidence is approximately 1 in 300, although the exact number is un-

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known secondary to underreporting [5]. The disease affects women more often than men and blacks more than whites, presumably because of a greater apocrine gland density [4, 6–8]. Predisposing factors include obesity, diabetes mellitus, perineal antiperspirants, poor hygiene, and smoking. A genetic predisposition has been suggested, with several reports of familial involvement suggesting a single gene transmission [9].

The pathophysiology of hidradenitis suppurativa appears to be follicular hyperkeratosis with plugging and dilatation of the follicle [1, 5]. Once occlusion of the gland occurs, microorganisms trapped by the keratin plug multiply, causing cellulitis and abscess formation. The distal aspect of the follicle dilates and eventually ruptures into the dermis, spreading the infection. With disease progression, spontaneous abscess drainage oc-

Figure 1. Inguinal and perineal hidradenitis suppurativa lesions. Multiple fistula tract openings are present in the inguinal and perineal regions.

Figure 2. Vulvar hidradenitis suppurativa. Fibrous vulvar mass lesion and numerous fistula openings are present.
curs through cutaneous tracts. The resulting wound does not heal completely, and recurring infections result. Chronic draining sinuses, intertwining cutaneous fistulae, and fibrosis result. The malodorous drainage carries with it a large social and psychological stigma that can lead to evading health care and to decaying interpersonal relationships [1].

Often, axillary hidradenitis suppurativa is mild, but perineal hidradenitis often is associated with frequent recurrence and poor outcomes. Many such patients have chronic abscesses and draining sinus tracts. In the chronic, severe state, distinguishing hidradenitis suppurativa from anal fistulae, lymphogranuloma venereum, granuloma inguinale, Crohn’s disease, chronic pyoderma gangrenosum, and carcinoma is difficult. A seeming association exists between Crohn’s disease and hidradenitis suppurativa, although no unifying mechanism for the diseases has been identified [10, 11]. In this case, the patient’s bowel history and the location and appearance of the lesions were suspicious for Crohn’s disease. However, the autopsy pathology was not consistent with Crohn’s disease. Though dermal fibrosis is not specific for hidradenitis suppurativa, these lesions most likely represent long-standing scarred lesions of hidradenitis suppurativa. That supposition is based on the 20-year history of draining axillary and vulvar “nodular pustules” and the absence of carcinoma and Crohn’s disease (a thorough examination of large and small bowel having shown no mucosal flattening or ulceration, strictures, granulomatous inflammation, or other features of Crohn’s disease).

Squamous cell carcinoma has been reported to occur in approximately 3.2% of perineal hidradenitis suppurativa patients, demonstrates male predominance, and tends to occur 20 to 40 years after initial development [12]. Though the appearance of our patient’s lesions was suspicious for carcinoma, none was found at autopsy.

Other reported medical complications from hidradenitis suppurativa include anemia, interstitial keratitis, osteomyelitis, fistulous communications to pelvic organs, and even death [13]. A Medline search from 1966 to 1997 revealed only one death reported to be caused directly by benign hidradenitis suppurativa [3]. That patient developed fistulas to the bladder, rectum, and peritoneum. Ultimately, he expired from peritonitis, pyelonephritis, and overwhelming sepsis after surgery. Lesser lesions can be treated with antibiotics, but the treatment of choice for advanced hidradenitis suppurativa is surgery, with skin grafts or healing by secondary intention.

Although the patient reported here also suffered from peritonitis and overwhelming sepsis at time of death, these disorders likely were secondary to her perforated sigmoid colon. In this case, the advanced stage of her disease, multisystem organ failure, and resultant depressed nutritional state precluded surgical management of her acute problem.

REFERENCES