Idiopathic pustular vulvitis

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Summary

A 31-year-old woman presented in the 23rd week of her third pregnancy with extremely painful pustular vulvitis, unresponsive to antibiotics. Although the histological findings were consistent with a diagnosis of hidradenitis suppurativa (HS), bridged comedones, the hallmark of this disease, were absent and there were no dermal sinuses. Incision and drainage of the pustules provided only temporary improvement, which was briefly maintained with oral clindamycin and topical steroids. After the birth, a course of isotretinoin produced almost total clearance, a response not typically found in HS. This patient’s condition may represent a variant of HS, and if so, it would be the first case report of de novo HS in pregnancy, but its clinical features and evolution differed so much from those in HS that the possibility of a previously unrecognised condition cannot be excluded.

Pustulosis of the vulva is an uncommon condition, many times confused with hidradenitis suppurativa. We present a case in which this distinction is discussed.

Report

A 31-year-old woman presented with a 2-day history of intense vulval pain and swelling. She found it difficult to walk because of the pain, and she was only comfortable lying down with no garments touching her vulva. She was in the 23rd week of her third pregnancy. She was in a monogamous relationship and denied any sexual contact for the previous 12 weeks. She had not changed any products for genital hygiene and her two previous pregnancies had been uneventful with vaginal deliveries at term. She had no relevant medical history; she had never had a sexually transmitted infection (STI) or any history of acne.

On physical examination, her pulse was 100 beats/min and her temperature was 37.2 ºC. She was in obvious distress with overwhelming vulval pain, which made a full vaginal examination difficult. The skin on both external labia majora was erythematous and studded with superficial pustules (Fig. 1). The mucosal surfaces were erythematous and oedematous, but showed no pustules. There were no signs of deep abscesses or fibrosis and no draining sinuses. No other skin abnormalities were present elsewhere. She had small tender inguinal lymph nodes.

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Figure 1 Clusters of pustules studded the labia majora. The labia minora and vagina were not affected and very few lesions affected the inguinal folds.
Routine laboratory tests, including blood sugar levels were all normal, except for a raised C-reactive protein (CRP) level of 108 mg/L (normal < 10). Cultures of blood and lesional pus were obtained, and the patient was started on intravenous flucloxacillin and benzylpenicillin. She required regular morphine for analgesia. The diagnostic possibilities at this time were staphylococcal folliculitis, hidradenitis suppurativa, Behçet’s disease, lymphogranuloma venereum, herpes simplex, granuloma inguinale, chancroid and localized pustular psoriasis.

By the third day of admission, there was no response to the antibiotics; on the advice of the infectious diseases team, these were changed at this point to cefotaxime, metronidazole, clindamycin and fluconazole. Culture of the pus grew normal skin flora only and blood cultures were negative. Other investigations included tests for various STIs, including VDRL, serology testing for human immunodeficiency virus, Treponema pallidum haemagglutination test and dark-field examination of the exudate to detect syphilis treponemes, serology testing and direct testing with scrapings from the base of a lesion (Tzanck test) for herpes simplex, and complement-fixation test for Chlamydia trachomatis. The results were all negative.

After a further 2 days, there was no improvement with the antibiotics and the diagnosis was still unclear. Consent was then obtained for further examination, incision, drainage and biopsy of pustules under spinal anaesthetic. Multiple pustules, each up to 10 mm in diameter, covered the labia majora. The deeper tissues were firm to the touch. The vagina, cervix and perianal areas were not affected. The roofs of the pustules could be promptly detached, leaving behind shiny, shallow erosions. There were no deep abscesses or sinus formation. Culture of a core of tissue for viruses, pyogenic bacteria, acid-fast bacilli and Haemophilus ducreyi had no growth.

An incisional biopsy taken from the lesional skin showed a moderately dense, predominantly deep dermal perivascular, interstitial and periappendageal mixed inflammatory cell infiltrate, composed of lymphocytes, neutrophils, plasma cells and occasional multinucleated giant cells, in association with vascular proliferation (Fig. 2a,b). Staining with Gram and periodic-acid–Schiff for bacteria and fungi was negative, as was the search for Donovan bodies using Warthin–Starry stain. These histological features were in keeping with granulation tissue, occurring in association with folliculitis with secondary rupture, as is seen in HS. There were no epidermal or dermal features compatible with pustular psoriasis.

The patient felt better after incision and drainage, but still required opiates. The cefotaxime and fluconazole were stopped 7 days later, at which stage her CRP was 9 mg/L. The patient remained on metronidazole and clindamycin for a total of 16 days. Local treatment included washing with saline, the application of ice packs and the use of topical mupirocin. One week following surgery, morphine was substituted with tramadol 100 mg three times daily and regular paracetamol. By the 26th week of gestation, the patient was able to walk, but new pustules continued to appear and...
were associated with considerable pain. At 35 weeks' gestation, the patient had developed a severely painful abscess, 30 mm in size on her left labium majus, which was incised and drained under spinal anaesthesia. The pus was sterile and the histology was very similar to that of the first sample. The patient underwent elective caesarean delivery at the 37th week of gestation to avoid further trauma to the vulva. A healthy female infant weighing 2.94 kg was delivered, who did not show any signs of opiate withdrawal.

The vulval lesions continued to recur postnatally, and the patient required regular tramadol, diclofenac and paracetamol for analgesia. At 5 weeks postpartum she was started on isotretinoin 1 mg/kg/day for 4 months. During this time, she used effective contraception and did not breast-feed. After 6 weeks of treatment, the active lesions resolved. One further lesion appeared on the right labium majus during treatment, which was injected with steroid. The patient also required further drainage of an abscess 9 months after treatment. Two years after completing the course of isotretinoin, she had no further lesions and her vulva had normal morphology, but she still had intense vulval pain on a very well defined point in the left labium major and remains under follow-up by the pain clinic.

At the end of all investigations and procedures, the proposed diagnosis for this patient was HS. This is a chronic relapsing inflammatory disease of apocrine-bearing skin, with a predilection for intertriginous areas, usually the axillary, inguinal, perianal and perineal areas. It presents as firm, erythematous nodules that evolve towards the formation of sinuses that discharge pus onto the skin surface. Ongoing chronic inflammation and recurrent episodes of exacerbation result in the formation of an intercommunicating system of sinus tracts in addition to severe fibrosis in the dermis and subcutaneous tissue. Previous studies have shown premenstrual exacerbations, improvement in pregnancy, and postpartum relapses. HS is controlled with difficulty. Multiple treatment regimens are available, including systemic antibiotics, infliximab, corticosteroids, incision and drainage, local excision, radiation and laser treatment. Clinical trials with large numbers of patients (evidence level 2) have shown marked improvement with the use of topical clindamycin and systemic tetracyclines. However, no single treatment has proved useful for all patients. Radical excision of the affected tissue is the most definitive treatment.

Retinoids have been used in the treatment of HS with limited therapeutic effect. Our patient's lesions responded dramatically to only one course of isotretinoin. She still has pain, but the vulva is morphologically normal, without scarring.

Although we cannot offer an alternative diagnosis to this woman's disease, we cannot subscribe to the diagnosis of HS either, because this case differs from HS in a number of ways. Her lesions had their onset in pregnancy, whereas HS improves during pregnancy. There were no firm, deep nodules, but numerous superficial pustules instead. Bridged comedones, the hallmark of HS, were absent and there were no dermal sinuses. The patient had very few lesions in the inguinoperineal area and none in the axillae, and atypically for HS, her condition responded dramatically to isotretinoin. This patient's condition might represent a new condition, waiting for further contributions to be better understood.

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References