Pyoderma Gangrenosum a Diagnosis not to Miss: Case Report

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ABSTRACT

Pyoderma gangrenosum is a very rare and aseptic ulcerative dermatosis with neutrophilic infiltration. It’s often associated with a chronic or systemic disease. The diagnosis is clinical. It’s a diagnosis of exclusion. It often mimics other diseases. The diagnosis is made later especially in gynecology-obstetric. That’s why we should not miss the diagnosis because the course is unpredictable and can be fatal.

Here we present a case of a 23 years old woman who presents a skin lesion in the right breast and a recurrence during pregnancy after the caesarean. The evolution was fatal, and the patient, unfortunately, died a few weeks later.

Keywords: Pyoderma gangrenosum, Pregnancy, Breast, Neutrophilic dermatosis

1. INTRODUCTION

Pyoderma gangrenosum is an unknown neutrophilic dermatosis. The incidence is about 3 to 10 million cases per year. It’s a diagnosis of exclusion. The clinical and histology symptoms are not specific. It’s barely associated with pregnancy and breast lesion. Few cases are reported in the literature.

2. CASE REPORT

The patient is a 23 years old breastfeeding woman, with four living children (4 births vaginally), and without any medical history. The patient was suffering a week before her admission from a painful edema in the right breast. She came to the maternity emergency nine months after her delivery because of a fast-growing breast abscess localized in the internal quadrant of the right breast. An ulcero-necrotic, painful lesion measuring 10cm x 4cm that occupies 50% of the breast with a renitent mass (Fig. 1) was found in the Admission exam. The breast ultrasound found a large collection with a thickened wall and heterogeneous content of the entire right breast. The biopsy didn’t find any malignant element. The management consists of surgical drainage of the abscess with the elimination of 500 mL of purulent fluid, with an intensive care lesion and an anti-microbial treatment. The diagnosis evocated at this moment was fasciitis necrosis. Following a good evolution, a skin graft was done with total remission (Fig. 2).
Nine months later, the patient was admitted for premature rupture of the membranes in 9 months pregnancy. Following this, a caesarean was realized because of premature rupture of the membranes. During her hospitalization, three days after the surgery, the patient had a pain in the post-operative wound with the apparition of pus (Fig. 2 & 3).

Fig. 1: The patient during her admission in the emergency

Fig. 2: the complication of the post-operative wound

A microbiological sample of the fluid was negative. The patient received antimicrobial treatment without any improvement. That’s why we made a skin biopsy which reveals an inflammation with neutrophilic dermal and hypodermal infiltration. The diagnosis of pyoderma gangrenosum was then established. The state of the patient got worse, and she was admitted to intensive care department. Different tests, in order to find an etiology, revealed a microangiopathy disease. The patient died a few days later.

3. DISCUSSION

Pyoderma gangrenosum is a rare, inflammatory neutrophilic dermatosis characterized by a noninfectious skin ulceration produced by Streptococcus(1). Brunsting was the first to use the term in 1930(1,2). It’s often located in lower limbs and abdomen(3). It appears in 0.63/100 000 cases, between 20 and 50 years old, mostly in women(3). It’s not only a skin disease but also a systemic disease. It’s associated with other diseases in 50 to 70% of time(1): inflammatory bowel disease in 15 to 48% (ulcerative colitis, Chron disease), joint and arthritis (seronegative arthritis, spondylitis of inflammatory bowel disease, rheumatoid arthritis), hematologic disease (myeloid leukemia, myelofibrosis, benign monoclonal gammopathy), other neutrophilic dermatosis (Sweet syndrome, subcorneal pustular dermatosis, Behcet), active hepatitis and episode of major depression suggest the diagnosis, systemic lupus erythematosus, cancer (breast, colon, prostate..)(3,4). In our case, the patient had a microangiopathy disease. Pathogenesis of pyoderma gangrenosum is still unelucidated. It might be due to an association between neutrophil dysfunction, genetic predisposition (a lot of family cases), immunologic dysfunction and infectious theory. However, the

presence of bacteria is probably a consequence of the superinfection of the ulceration\(^4\). The diagnosis is clinical. First of all, it appears as a painful edematous, erythematous lesion with inflammatory characteristics that expands rapidly and develops into a painful ulcer with mucopurulent or bloody exudate and septicemic symptoms in aseptic lesion\(^4\). It may happen spontaneously, after surgery (6 to 14 days) or due to injury\(^1(2)\). An aggressive surgery may worsen the lesion; it’s the pathergy effect\(^5\). The microbiological samples are often negative. The histology is not specific. The skin biopsies find an inflammatory infiltrate with neutrophilic dermal and hypodermal infiltration without any germ or infection signs with Central epidemics necrosis\(^6,7\). As our patient’s Immunohistochemistry shows perivascular deposit\(^4\). It can be underdiagnosed. Indeed, it can mimic necrotizing fasciitis (a lot of cases were described), surgical wound infection, bacterial necrotizing dermo-hypodermitis, cutaneous infection, sepsis, sweet syndrome ulcerative form, malignant lesion, vascular disorder\(^9\). We had the same problem with our patient, firstly we considered the breast lesion as a fasciitis necrosis and then the wound lesion as a superinfection of it. The diagnosis of pyoderma gangrenosum is a diagnosis of exclusion. In fact, we must in a first time eliminate the other causes of similar skin ulceration\(^2\), a wound “infection” or infection cellulitis\(^2\). The diagnosis is based on the clinical grounds: if the lesion decreases after systemic corticoid therapy\(^5\). The absence of germs in the different test, the no-amelioration after anti-microbial treatment\(^2,4\). There is no gold standard therapy. The treatment is more effective if it begins quickly. The therapy depends on the rapidity of development of the lesion, the size, the depth, the appearance of a new lesion and the presence of systemic disease\(^2\). The treatment should be multidisciplinary. The treatment of pyoderma gangrenoso is based on medical treatment. If the lesion is superficial or localized, we can only use a topical treatment like corticoid, tacrolimus, and isotretinoin but it has teratogenic effects. In the case of a severe form, the main treatment is immunosuppressive by using cyclosporine, oral or intravenous corticoid with an initial dose of 1 to 2 mg per kg per day but the duration is not codified (few weeks to few months), azathioprine, cyclophosphamide, plasmapheresis, no sterooidal anti-inflammatory\(^1\). New treatment can be used like anti-TNF alpha, growth factors. In the case of a resistant form, we may need mycophenolate mofetil, GM-CSF, autologous transplant and cultured cells. But surgical treatment must be very careful. It can be unnecessary, deleterious and have poor aesthetic effects. The surgery and debridement should not be aggressive because it will worsen the lesion and increase the morbimortality\(^3\). Indeed, we may need split skin graft\(^4\). The course is unpredictable\(^3\). Generally, the evolution is good even with a large lesion. The recovery is slow. But, the mortality is about 30%\(^3\).

In the literature, we only find 16 cases of pyoderma gangrenosum and pregnancy\(^8\). 9 cases are in postnatal, 6 cases are associated with other diseases and 7 cases in a recent wound (pathery effect) like in our case. In 10 cases, pregnancy is the only cause of pyoderma. It shows that the pregnancy might be the only contributing or trigger factor\(^9,10\). Indeed, gestation is a state of humoral and cellular immunosuppression. It increases different coagulation factor and influences the relationship between immune and endocrine systems leading to an inflammatory cascade, as the origin of pyoderma. Another theory incriminates G-CSF (granulocyte colony stimulating factor).

Pyoderma gangrenosum doesn’t have any effect on the pregnancy and the fetus\(^1,10\). At the same time, pregnancy has no influence in pyoderma gangrenosum. It doesn’t slow or stops the progression of healing. For example, some case of abortion didn’t stop the evolution of pyoderma gangrenosum\(^8,9\). The problem with breast localization is that we should first eliminate malignant disease, and then make a difference between paraneoplastic pyoderma and the consequence of injury due to breastfeeding like our patient or post-operative\(^4\).

4. CONCLUSION

Thus, although the pyoderma gangrenosum is clinically characteristic, it remains an enigma with regard to its etiopathogenesis. There are various clinical and histological variants of the disease. Criteria have been proposed to diagnose the pyoderma gangrenosum.
REFERENCES


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