

CASE REPORT

Pyoderma vegetans of the posterior area of the neck: case presentation

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Abstract

Pyoderma vegetans is a rare disease characterized by the presence of vegetant exudative, pustular and erythematous vesiculobullous plaque usually located in the inguinal area and axillary fold. Etiology of pyoderma vegetans is unknown but it is often associated with bacterial infections in immunocompromised patients. Main histopathological characteristics of pyoderma crops are pseudoepitheliomatous hyperplasia and subepidermal, intraepidermal neutrophilic or eosinophilic microabscesses. It is well known that these lesions are commonly associated with colonic inflammatory disease such as ulcerative colitis and Crohn's disease. Not available standard treatment for pyoderma vegetans, although the use of antibiotic therapy was often used with variable results. Standard first-line therapy is the systemic steroids yet. We perform excision of the lesion of the posterior area of the neck with application of the free split-thickness skin graft after 48 hours postoperatively. In this paper, we present a case of pyoderma vegetans with unusual location without associating colonic lesions and a review of literature related to therapeutic and diagnostic problems of this disease.

Keywords: pyoderma vegetans, pseudoepitheliomatous hyperplasia, inflammatory dermatosis.

Introduction

Pyoderma vegetans is a chronic inflammatory dermatosis very rare. It is characterized by the presence of verrucous plaques, large exudative and well-defined borders [1]. Pyoderma vegetans usually affects middle-aged men and is commonly associated with ulcerative colitis, which is difficult to treat [2]. Other diseases associated with this entity include cutaneous T-cell lymphoma, primary immunodeficiency, chronic myeloid leukemia, alcoholism, HIV (human immunodeficiency virus) infection and nutritional deficiencies [2–4]. However, pyoderma vegetans been reported in patients without other associated diseases [2, 5]. Major histopathological features are pseudoepitheliomatous hyperplasia and subepidermal, intraepidermal neutrophilic or eosinophilic microabscesses [6].

Aim

We describe the case of a 72-year-old man with long evolving pyoderma vegetans without any history of an immunocompromised condition. Our patient is unique in its presentation of pyoderma vegetans of posterior area of the neck with excision of the lesion of the posterior area of the neck and application of the free split-thickness skin graft after 48 hours.

Case report

D.I., 72-year-old male patient was hospitalized in Second Surgical Clinic, Emergency County Hospital, Craiova, Romania, on 17.01.2011, FO 2761 for neck pain and the appearance of a tumor at this level ago about two years.

On physical examination, he displayed the presence of a tumor vegetans in the neck region with dimensions of 15/8 cm, uneven surface, edges elevated, with many areas fluctuant, yellowish crusts on the surface, erythematous edges involving almost the entire region of the neck slightly sensitive on palpation (Figure 1).

On admission laboratory tests revealed: hemoglobin 13.6 g/dL → 10.5 g/dL → 10.8 g/dL; white blood count 9600/mm³ → 9200/mm³; glycemia 147 mg% → 123 mg% → 101 mg% → 109 mg%; Quick time 101%; INR (international normalized ratio) 1; urine analysis normal: urea 39 mg%, creatinine 0.81 mg%; total bilirubine 0.57 mg%; ALAT (alanine aminotransferase) 15 IU; SGOT (serum glutamic oxaloacetic transaminase) 15 IU; sedimentation rate 30 mm/h.

ECG (electrocardiography): sinus rhythm, QRS axis normal, without abnormal repolarization.

Chest X-ray: no active pleuro-pulmonary lesions.

Colonoscopy: rectum and colon mucosa up to the ileocecal valve for normal endoscopic appearance without localized processes.

Abdominal ultrasound (US): liver without localized processes, gallbladder without stones images. Right and left kidney normal. Spleen and urinary bladder were normal ultrasound appearance.

Evaluation of skin biopsy revealed pseudoepitheliomatous hyperplasia, fibrosis, vascular proliferation and

predominantly mononuclear infiltrate with neutrophils in the dermis.

The surgical intervention and lesion excision is practiced by healthy tissue (Figure 2) followed within 48 hours with application of the free split-thickness skin graft in the remaining area (Figure 3).



Figure 1 – Preoperative tumor appearance in the posterior region of the neck. Yellowish crusts on the surface are observed, erythematous edges involving almost the entire posterior region of the neck.



Figure 2 – The surgical intervention and tumor excision is practiced by healthy tissue.



Figure 3 – At 48 hours after surgery for removal of tumor formation, are free split-thickness skin graft practice.

Histopathological examination revealed: intraepidermal abscess (Figure 4), multinucleated giant cells (Figure 5), intradermal microabscesses. In the epidermis was found the presence of neutrophils and fibrin lined ulceration and squamous epithelial hyperplasia (Figure 6). Dermal compartment was found the presence of edema, fibrosis, vascular proliferation and neutrophilic infiltration in several vascular walls without association fibrinoid necrosis and plasma cells, occasional neutrophils predominant inflammatory dermis. Based on clinical examination and histopathological findings the diagnosis was pyoderma vegetans of neck.

After removal of tumor developed in the posterior area of the neck was obtained operative piece measuring 15/8 cm composed of skin, subcutaneous tela, nuchal fascia. Epidermal surface was found the presence of numerous yellowish crusts and upon section were found numerous intraepidermal abscesses and intradermal edema.

After removal of the lesion, the area remaining after surgery, the neck region was covered with free split-thickness skin graft after 48 hours postoperatively.

Surgical specimen was fixed in 10% formalin solution for three weeks. After fixation were performed 1 cm thick sections through operative piece and were processed in paraffin blocks 12 pieces from different parts of the lesion using standard histological techniques. Five μ m sections were stained with Hematoxylin–Eosin (HE), Gömöri technique for reticulin, and orcein method for elastin. Microscopic examination was performed using Nikon microscope.

Immunohistochemical examination was performed on paraffin blocks containing tumor formation eradicated of the posterior area of neck region. Immunohistochemical technique was the standard processing.

Immunohistochemical examination revealed: CD20-positive B-lymphocytes, CD68-positive giant macrophages, CD45RO-positive T-lymphocytes, EMA (epithelial membrane antigen) positive plasmocytes.

Postoperative evolution was favorable without incident. The patient was discharged after 12 days cured surgically.

Then reviewed after six months (Figure 7), nine months and one year progress was favorable, the patient presenting with clinical status improved.

Colonoscopy, chest X-ray, abdominal US performed preoperatively revealed normal relations. Surgical intervention and the tumor were removed from the neck region to healthy tissue, followed by application of the free split-thickness skin graft at two days postoperatively.

Dimensions of operative piece was 15/8 cm, with irregular surface, firm flesh sometimes fluctuant, yellowish crusts on the surface. Histological examination revealed pseudoepitheliomatous hyperplasia, abscesses dermal and epidermal and dermal neutrophilia, multinucleated giant cells, without the presence of granulomas.

Pathognomonic lesions in pyoderma vegetans have two important characteristics: pseudoepitheliomatous hyperplasia, numerous abscesses in both the dermis and epidermis as hyperplastic.

Abscesses may consist of neutrophils or eosinophils.

Microscopic histopathological examination of operative piece revealed: intraepidermal abscess, intradermal microabscess, and giant cell granuloma reaction. In the epidermis was found the presence of neutrophils and fibrin lined ulceration and squamous epithelial hyperplasia. Dermal compartment was found the presence of edema, fibrosis, vascular proliferation and neutrophilic infiltration in several vascular walls without association fibrinoid necrosis and plasma cells, occasional neutrophils predominant inflammatory dermis.

Immunohistochemistry can be used to determine the presence of CD20 on cells in tissue histological sections. CD20 can be very useful for the diagnosis of B-cell lymphomas and leukemias.

CD20 is a non-glycated transmembrane protein expressed on precursor B-cells and mature B-cells but absent in plasma cells upon differentiation. Once the

CD20 is not stained histiocytes or plasma cells and was not detected in T-cell malignancies, is considered an important marker for B-cell lymphomas.

In our case, CD20 was present in B-cells from sections made of operative piece (Figure 8).

There are several isoforms of CD45: CD45RA,

CD45RB, CD45RC, CD45RAB, CD45RAC, CD45RBC, CD45RO, CD45R. CD45RA is located on naïve T-cells and CD45RO is located on memory T-cells. CD45RO is recognized as a protein of 180 kDa. In our case, CD45RO was revealed on histological sections and was present in T-cells (Figure 9).

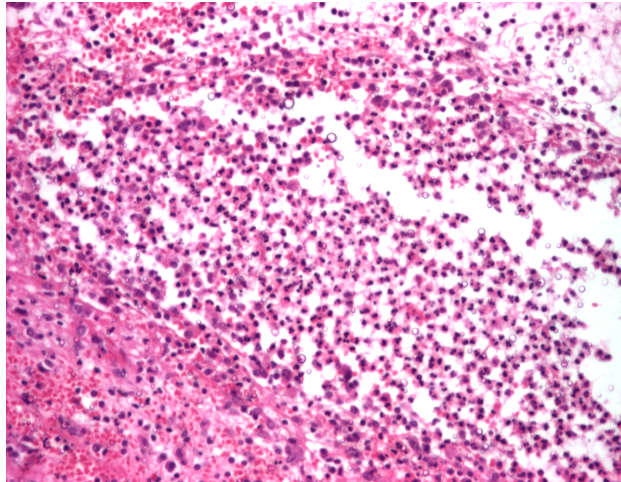


Figure 4 – Intraepidermal abscess. HE staining, ×100.

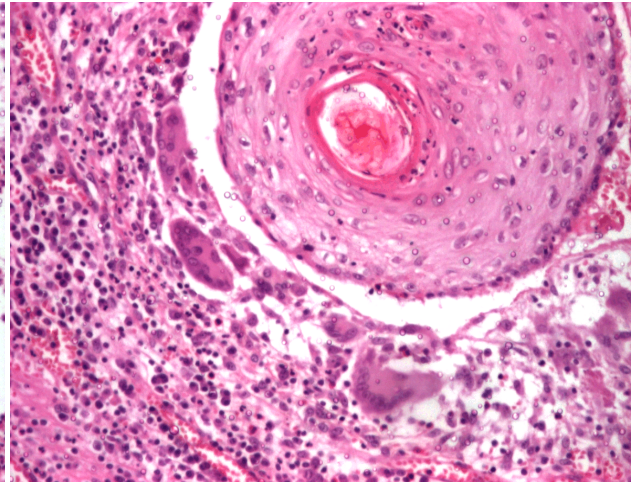


Figure 5 – Multinucleated giant cells. HE staining, ×100.

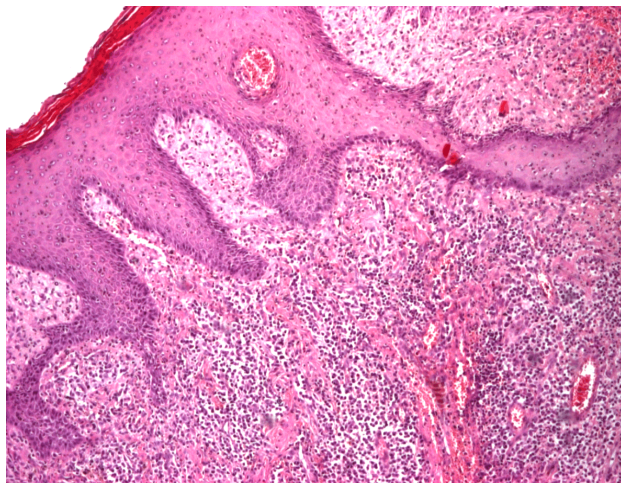


Figure 6 – Pseudoepitheliomatous hyperplasia. HE staining, ×40.



Figure 7 – The posterior area of the neck appearance at three months postoperatively, showing healing of the lesion.

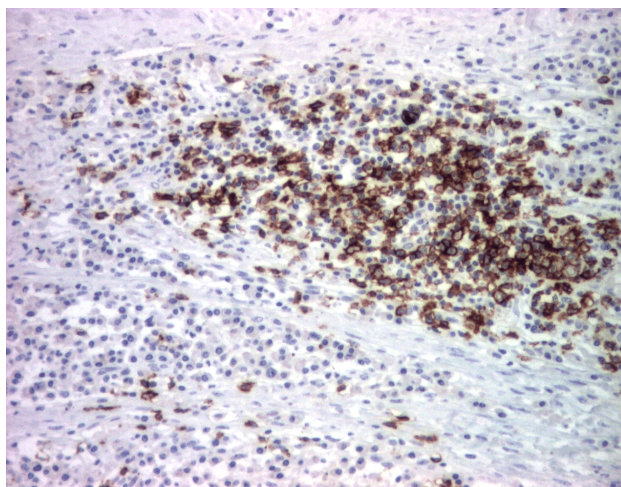


Figure 8 – CD20-positive B-lymphocytes. Immunostaining with CD20 antibody, ×100.

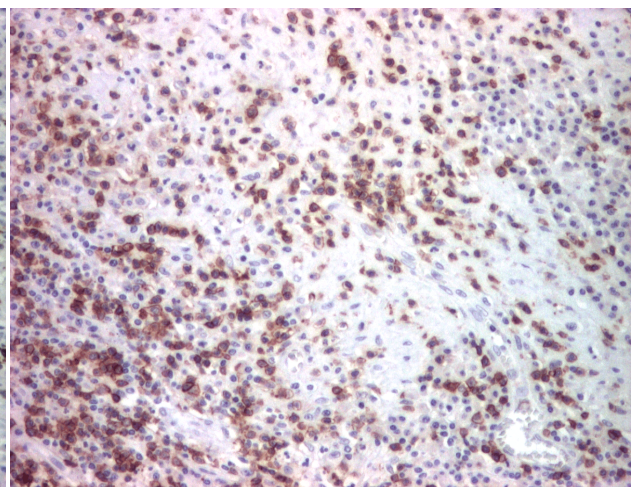


Figure 9 – CD45RO-positive T-lymphocytes. Immunostaining with CD45RO antibody, ×100.

Immunohistochemistry can be used to identify CD68 that is found in cytoplasmic granules of blood cells. Is particularly helpful as a marker of various macrophage cell line spleen, lamina propria of the intestine, lung alveoli and bone marrow, including histiocytes, giant cells, Kupffer cells and osteoclasts.

On histological sections, in our case was the presence of CD68-positive giant macrophage and immunohisto-

chemical examination with CD68 staining revealed no granuloma formation (Figure 10).

EMA is a glycoprotein found in secretory cells of mammary secretion. EMA antibody is helpful in the diagnosis of extramammary Paget's disease, neoplasms of eccrine and sebaceous carcinomas. In our case, EMA was found in the plasmocytes (Figure 11).

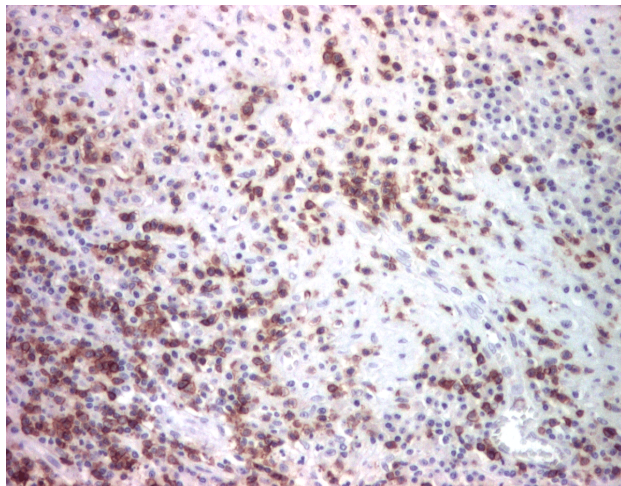


Figure 10 – CD68-positive giant macrophages. Immunostaining with CD68 antibody, $\times 100$.

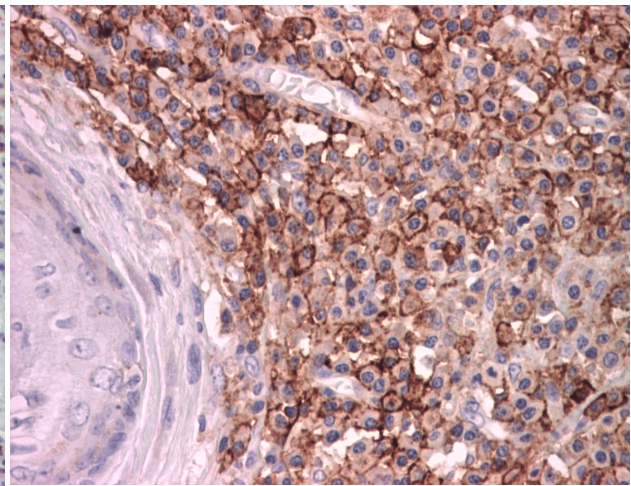


Figure 11 – EMA-positive plasmocytes. Immunostaining with EMA antibody, $\times 200$.

Discussion

Pyoderma vegetans is a very rare chronic inflammatory dermatosis and also named blastomycosis-like pyoderma. When mucosal lesions are observed it is referred to as “pyostomatitis vegetans”. The literature was reported that the disease coexists with a variety of diseases including ulcerative colitis, diffuse T-cell lymphoma, chronic alcoholism, HIV infection and chronic myeloid leukemia [3, 5, 7–12] but disease has been reported in other patients without other diseases [5, 13, 14] as in the case reported by us.

The most frequent microbial agents isolated were β -hemolytic staphylococci and streptococci. Other suspected microbial agents were *Klebsiella*, *Bacteroides*, *Enterococcus*, *Pseudomonas aeruginosa* and *Corynebacterium* [3–5, 15, 16]. A case associated with *Trichophyton mentagrophytes* has also been reported [14]. Some authors believe that microbial agent is not responsible for pyoderma vegetans primary and secondary infection develops an immune dysfunction [5, 14]. In support of this theory, in some cases, serum immunoglobulin levels were low and phagocytic dysfunction was observed [4, 8–10, 13, 14].

Pyoderma vegetans thought that was the result of bacterial invasion in immunocompromised patients [1]. Diagnosis of the disease depends on five criteria were proposed by Su *et al.* [13]: large plaques with multiple pustules and elevated edges, pseudoepitheliomatous hyperplasia with abscesses in biopsy tissue growth at least one pathogenic bacteria, negative culture for fungi, for atypical mycobacteria and *Mycobacterium tuberculosis* and normal levels of bromide.

Should also be excluded skin tuberculosis skin, pyo-

derma gangrenosum, Sweet's syndrome and squamous cell carcinoma [1–3].

Pyoderma vegetans to differentiate from other diseases, wound assessment, the secretions and culture for *M. tuberculosis* and direct immunofluorescence study will be conducted to exclude pemphigus vegetans. Association of pyoderma vegetans with ulcerative colitis is well known but rarely reported in the literature [2, 3, 6, 7, 17, 18]. However, pyoderma vegetans is considered a marker of inflammatory colonic disease and a presumptive diagnosis of pyoderma vegetans induce gastrointestinal completes an investigation. In our case, colonoscopy performed up to the ileocecal valve revealed normal relations.

Immunohistochemistry is useful in making the differential diagnosis of a range of diseases. We found immunohistochemical examination: CD20-positive B-lymphocytes, CD68-positive giant macrophages, CD45RO-positive T-lymphocytes and EMA-positive plasmocytes. CD20 can be very useful for the diagnosis of B-cell lymphomas and leukemias. Sometimes, CD20-positive cells can be found in patients with Hodgkin's disease, myeloma and thymoma [19]. CD20 antigen reacts with cell membrane present in B-cells. This antibody recognizes strongly Reed–Sternberg cells, predominant Hodgkin's disease. The B-lymphocyte antigen is expressed on the surface of B-cells from pro-B-stage and gradually increasing concentration to adulthood [20].

CD45 was originally called leukocyte common antigen [21]. This antigen is expressed on virtually all cells of hematopoietic origin, except erythrocytes and plasma cells, which assist the activation of these cells. This antibody stained cells in lymphomas, B-cell chronic lymphocytic leukemia, “hairy” cell leukemia, non-lymphocytic leukemia.

CD68 is a glycoprotein that is expressed on monocyte-macrophage line [22, 23]. Presence in macrophages is useful for diagnosing malignant conditions where the presence of this cell such as the malignant histiocytosis, lymphoma and Gaucher's histiocytosis [19, 24]. CD68 can also be helpful in differentiating malignant histiocytosis other pleomorphic sarcomas.

Although many treatments have been tried for pyoderma vegetans, the pathologic entity is very difficult to treat. The treatment of pyoderma vegetans focuses on control of underlying disease and the lesions regressed after management of bowel disease with Sulfasalazine, Mesalazine or subtotal colectomy [1–3]. Despite bacterial etiology of pyoderma vegetans lesions, antibiotics alone are not an effective treatment [13, 15]. Topical application of Disodium Cromoglycate has been used in the treatment of pyoderma vegetans, which was refractory to antibiotic treatment [25]. Standard therapy is represented by systemic steroids at a dose of 0.5–1 mg/kg. Response to systemic steroids is usually very good [15, 26]. Many other therapeutic modalities including: Dapsone, Azathioprine, Cyclosporine and laser debridement have been reported and accepted as secondary therapy lines [1, 2, 18, 26]. Despite these therapies, total colectomy filling Tacrolimus has also been reported to be beneficial in eradicating lesions [18].

Carrera *et al.* [1] also reported a patient with pyoderma associated with psoriasis that was successfully treated with Etanercept. Another patient with ulcerative colitis associated with pyoderma vegetans was treated with Sulfasalazine for ulcerative colitis and skin lesions was treated with steroids and antibiotics.

Pyoderma vegetans has good prognosis after therapy. Evolution of the pyoderma vegetans tends to follow the progression of underlying bowel disease [15].

Conclusions

It is important to recognize this vegetans skin lesion so that a correct diagnosis can lead to the discovery of inflammatory bowel disease. In our case, surgical treatment, which consisted of complete excision of tumor formation of the posterior area of the neck, with application of the free split-thickness skin graft after 48 hours postoperatively leading to lesion healing.

Conflict of interests

The authors declare that they have no conflict of interests.

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