

# Aseptic Systemic Abscesses

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## Abstract

*The syndrome of aseptic systemic abscesses is now a well-defined entity inside the autoinflammatory disorders; it is a rare disease -as about only 30 cases have been documented in France since 1999- that affects mainly young adults. This syndrome is characterized by recurrent attacks of fever and deep abscess-like collections most frequently localized in the abdomen. Blood markers of inflammation and polymorphonuclear neutrophils are elevated. All researches for a pathogen including PCR with universal and specific probes remain negative. On pathologic examination, aseptic abscesses are made of a core of polymorphonuclear leukocytes more or less altered surrounded by palisading histiocytes and sometimes giant cells. Antibiotics fail to cure the patients that improve dramatically with corticosteroids and immunosuppressive drugs. Aseptic abscesses may be either isolated or associated with an underlying condition such as relapsing polychondritis or inflammatory bowel disease that they may reveal for several years. A neutrophilic dermatosis like pyoderma gangrenosum may also be observed. A familial history of granulomatous disorder is available in a few cases.*

## Keywords

aseptic abscess –autoinflammatory disorder – corticosteroid – Crohn disease – inflammatory bowel disease – neutrophilic dermatosis – polymorphonuclear neutrophil – pyoderma gangrenosum – Sweet syndrome.

## Disease name and synonyms

The first case of aseptic systemic abscesses has been published in 1995 (André *et al.*, 1995). The terms “aseptic abscesses syndrome”, “corticosteroid-sensitive aseptic abscesses” or “disseminated aseptic abscesses” may be also used.

## Definition

The aseptic abscesses syndrome is an inflammatory condition characterized by deep well defined sterile collections of polymorphonuclear neutrophils, usually associated with pain, high-grade fever and leukocytosis. The evolution of abscesses is not

influenced by antibiotic therapies; on the contrary, they are highly sensitive to corticosteroid therapy (André *et al.*, 1998). Aseptic abscesses are closely related to neutrophilic dermatoses and inflammatory bowel diseases (André *et al.*, 1997; André *et al.*, 2001; André *et al.*, 2005).

## Diagnosis

The diagnosis of aseptic abscesses syndrome must be raised when a patient develops deep abscesses and when extensive and repeated investigations fail to detect any pathogen. In this condition, an underlying inflammatory bowel disease or a neutrophilic dermatosis may be

suggestive of aseptic abscesses. Procalcitonin level may be useful to establish the differential diagnosis of infectious versus aseptic abscesses (Delevaux *et al.*, 2003).

### Differential diagnosis

An infection due to viruses, bacteria or other organisms must be ruled out carefully. Patients should be tested for HIV. Particular attention must be paid to infective endocarditis. Fungi, parasites, *Mycobacterium tuberculosis* or nontuberculous mycobacteria, *Chlamydia trachomatis*, *Bartonella henselae* and *Yersinia* infections may share the same pathological findings (Kémény *et al.*, 1999) that aseptic abscesses. A [chronic granulomatous disease](#) characterized by pyogenic or fungal recurrent infections with granulomatous formation can be excluded by normal granulocyte function tests and negative researches for pathogens. The differential diagnosis includes also inflammatory conditions such as [Wegener's granulomatosis](#) or [Weber-Christian disease](#) and malignancies such as [Hodgkin's disease](#).

### Etiology

The cause of aseptic systemic abscesses is unknown. Although there is no obvious Mendelian inheritance, patients with aseptic abscesses syndrome have often familial history of granulomatous disorder such as [Crohn's disease](#) sometimes associated with cystic acne, in 15% of the cases.

### Frequency

A national study (Société Nationale Française de Médecine Interne) that was started in 1999 has permitted to identify and to follow-up more than 30 cases of aseptic abscesses up to now.

### Clinical description

Aseptic abscesses syndrome affects young adults of both sexes with a mean age at onset of 30 years (André *et al.*, 2002). Aseptic abscesses are mainly located in the abdomen: they involve by order of decreasing frequency: spleen, abdominal lymph nodes, liver and pancreas. Other organs outside of the abdomen such as lung, brain, muscle or pharynx may be concerned. The illness typically begins with abdominal discomfort and eventually diarrhoea, weight loss and low-grade fever for several weeks. At the time of presentation, the patients usually have high-grade fever but it is important to note that they have no deterioration in their haemodynamic status. They often have abdominal pain and tenderness without rigidity. Additional symptoms include arthralgia or arthritis, myalgia and mouth ulcers. Cutaneous involvement may be noted as [Sweet's syndrome](#), [pyoderma gangrenosum](#), neutrophilic

pustulosis or acne. Clinical features of inflammatory bowel disease are present in more than half of the cases. The abscesses usually precede the diagnosis of inflammatory bowel disease or are concomitant, but may also be subsequent to it.

Laboratory data show marked leukocytosis reaching up to 48,000/mm<sup>3</sup> with predominantly mature polymorphonuclear leukocytes and sometimes a mild or frank anemia; erythrocyte sedimentation rate and C-reactive protein are elevated. Liver enzymes may be mildly to moderately high. Autoantibodies are negative except for perinuclear antineutrophil cytoplasmic antibodies without antimyeloperoxidase specificity in a few patients.

Ultrasound and CT scan demonstrate multiple focal hypoechogenic or hypodense lesions in the organs involved. At this stage, a needle biopsy or an exploratory laparotomy is often performed for further investigations. Histopathologic examination of aseptic abscesses evidences a central suppuration containing more or less altered polymorphonuclear leukocytes surrounded by palisading histiocytes and sometimes giant cells.

### Management

A careful exclusion of another etiology and especially an infectious cause is mandatory. Intravenous or oral corticosteroids (prednisone 1mg/kg/day) achieve a rapid improvement. However, steroid-sparing drugs such as azathioprine or cyclophosphamide may be useful when high doses of corticosteroids are required (André *et al.*, 2003). Aseptic abscesses relapse in more than half of the cases at the same place or in another organ.

### Unresolved questions

Neutrophilic dermatoses are probably closely related to aseptic abscesses and share possibly the same spectrum: some authors reported deep sterile collections of polymorphonuclear neutrophils occurring during [Sweet's syndrome](#) or [pyoderma gangrenosum](#). Interestingly, a familial and systemic form of pyoderma gangrenosum with an autosomal dominant inheritance called PAPA syndrome (for Pyogenic sterile Arthritis, Pyoderma gangrenosum, and Acne) has been described (Lindor *et al.*, 1997). Four patients with PAPA syndrome developed sterile abscesses at sites of parenteral injections, evocative of a pathergy phenomenon. These clues suggest that neutrophilic dermatoses should rather be considered as a superficial feature of a systemic disorder including aseptic abscesses.

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