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Case Report

# Acute Gangrenous Appendicitis in A 70-Year-Old Patient with Multiple Myeloma

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## Introduction

Nowadays, because of the plethora of available literature, the correlation between acute appendicitis and hematological conditions due to immunosuppression and leukopenia is arrant. More precisely, 30% of all adult patients with hematological disorders will present with a gastrointestinal tract infection [1,2], while in the pediatric population in the presence of diseases like acute leukemia and lymphoma, the percentage of the effect reaches up to 1,5% [3]. An essentially interesting case report associated with the antecedently mentioned facts is the multiple myeloma which, usually appeared approximately in the 70th decade of life [4]. In the following report, we present a case of a patient with acute gangrenous appendicitis, accompanied by neutropenia because of the multiple myeloma. Because of the special nature of the disease, the physician has to elucidate if this situation incorporates a simple infection, due to the immunosuppression, when the cecum or the appendix is involved, and if there is an extramedullary localization of the disease either in the ileocecal area or the appendix. According to the previously mentioned details, the physician will be able to decide for each patient individually, based on medical evidence, followed by standard guidelines.

### Case Presentation

A 70-year-old patient presented to the surgical emergency department of the University General Hospital of Thessaloniki AHEPA, complaining of lower abdominal pain with a Blumberg positive sign on clinical examination. Diving into the personal history presence of multiple myeloma disease (known for 7 years), was discovered,while the patient was under follow-up and treatment of the disease.

The laboratory testing revealed 2000 White Blood Cells (WBCs) with neutrophils reaching up to 50% and 1000 k/mg neutrophils. In addition to the personal history, the patient referred as a recent intervention of tape placement due to uterine prolapse, a fact, which made the differential diagnosis even more difficult. Because of the complexity of the history, the patient underwent a series of imaging studies. The abdominal Computed Tomography (CT) revealed acute appendicitis, making due for a remark distention of the appendix and surrounding tissue inflammation. The imagistic findings of CT were not associated with the recent intervention, related to the uterine prolapse, but described an intraluminal tumor formation near the appendiceal base, which was not given, prominence to the plasmacytoma, as the image did

not have any characteristic of a soft tissue evidence. This tumor that was identified on the abdominal CT was attributed either to coprolith or felt. However, the question could be clarified only by histopathological examination. Furthermore, there was no description of an inflammatory reaction to the cecum.

Because of the differential dilemma due to the imaging findings (appendicitis, intraluminal tumor formation) and mainly due to the clinical image, the patient was taken to the operating room. The operation initially started via laparoscopy, however, it was converted via a middle-line incision to open surgery, due to the appendix location, which was in the greater omentum, and the appendiceal base, which was precarious. More specifically, the inflammatory area was significatly thickened, so its preparation was difficult. A standard appendectomy was performed without burial of the stump, something which was unavoidable due to the increased thickness of the inflamed area. Following the process, at the end of the procedure, an omental patch was placed above the stump, to achieve a higher percentage of safety. The patient recovered smoothly from anesthesia and was transported to the surgical department. She had an uneventful postoperative course. On the laboratory testing, WBCs showed no further decline, while other inflammatory markers subsided. The patient was discharged 48 hours postoperatively with the recommendation of a meeting with a hematologist. The histopathological examination revealed gangrenous appendicitis with the presence of coprolith. One year after the surgery, the patient has no symptoms related to the abdominal area.

## Discussion

Acute appendicitis has an increased frequency of appearance in patients suffering from hematological diseases. This condition is mainly noticed in children, however the epidemiological data changes when leukopenia and immunosuppression due to hematological conditions are involved [2,3]. One of these plights is the multiple myeloma, which is mainly observed after the 70th decade of life onwards [4]. Returning to the part of the scenario that refers to the infection, it is worth mentioning, that the bacterial infection importantly increases the rate of morbidity and mortality in patients suffering from multiple myeloma [5]. The most frequent infections are pneumonia, osteomyelitis, pyelonephritis and less commonly, intra-abdoominal infections [5]. Bacterial infections are a quite frequent cause of mortality and morbidity in the previously mentioned patient's category. On the one hand, bacterial infections are noted on solid organs, while on the other hand, intra-abdominal infections are not rare [2,5]. After all, it is acknowledged that Gram-negative bacterial infections have higher frequency rates in patients with neutropenia and the change of the infection's microbiological profile about those of the community, as well [6]. A particular circumstance is neutropenic enterocolitis or more precisely cecitis, known as either typhlitis or ileocecal syndrome [7].

In general, this condition is usually treated conservatively. Initially, by broad-spectrum antibiotic administration, fluids and electrolytes balance are administrated while in certain cases the injection or administration of cortisone and hematopoietic growth factors are also used [8,9]. An intriguing case in among these patients is cecitis, in which we may find inflammation expansion towards the appendix [7,10]. Cecitis or thyphlitis as it is also known is usually treated conservatively and due to the particularity of the disease, plenty of questions are raised regarding the differential diagnosis. The major question that has to be answered, is when there is a real appendiceal infection and when the surgical treatment via exploratory laparotomy is the only option [11,12]. At the time of differential diagnosis, one of the main problems that appears is the occasion in which there is an inflammatory reaction in the appendix and the surrounding area [10-13]. Taking this into consideration, the decision for surgical invasion will be based on the clinical appearence, as well as, the imaging finding (free intraperitoneal air, intraperitoneal fluid diffusion, inflammation of the cecal base) [7]. Furthermore, it is essential to mention that in multiple myeloma the physician-surgeon, must take into consideration the possibility of extramedullary localization of the disease, even if, according to the data the gastrointestinal occurrence is rare, especially in the appendix [14,15]. The extramedullary localization of the disease constitutes approximately 3% of the entire plasmacyte's neoplasias category [16]. Of those 80% are tumors observed on the head and neck and only 5% are composed of gastrointestinal solid tumors as extramedullary localization [17] [18]. Talking about localization, it is impoortant to mention some of the most extremely rare anatomical areas of identification, the large intestine, as well as, the rectum, especially the iileocecal area [19,20] and the appendix [15].

Thinking about the imaging part and the finding, we notice that computed tomography the plasmacytes, appear as soft tissue tumors [14,15], while in the necrotic enterocolitis or cecitis, the point findings are the distention of the intestine with mucosal lesion and the increased wall thickness [7]. Likewise, the appendix in the case of patients with neutropenia, appears enlarged with increased thickness of the wall accompanied by mucosal damage [10]. Coprolith or felt can also exist. The existence of a certain corresponding tumor raises the suspicion of the extramedullary localization of the disease, on the wider anatomical area of the ileocecal valve, essentially when it is imagistically combined, with a soft tissue texture image [7,10,14]. This fact is due to the pathophysiological mechanism of enterocolitis, which is involved in the process of mucosal damage and ischemia, as well as, reduced lymphatic drainage [7]. In addition, quite an important field is that,

of radiotherapy and chemotherapy, in which plasmacytes present high sensitivity [21]. Unfortunately, this perspective cannot be applied in the abdominal region, but only to the head, neck and thorax [14,16]. The gold standard in the anatomical area of the abdomen is the excision of the tumor, even with minimal or limited mucosal resection by endoscopy (EMR) [22]. Dismally, due to the anatomy of the appendix, either biopsy or EMR is forbidden. According to the previously mentioned data, appendectomy is the only available pathway, especially when the occurrence frequency of plasmacytoma is extremely rare. It is worth noting that another phenomenon is observed in the rest of the appendiceal tumors, where a further decision is made, following the histopathological results and imagistic findings. However, plasmacytoma, as well as, the rest of the appendiceal tumors present the same clinical image on patients, this of a right lower quadrant pain, and even with the clinical mage of acute appendicitis [14,15].

One of the most essential questions raised from this discussion is if the appendicectomy will be interpreted immediately or in a later phase in patients with aplastic crisis, especially those who underwent treatment with bone marrow transplantation [11,12]. Uthayanan et al support the theory of second-phase treatment when the bone marrow takes over with or without the use of growth factors. The index which is taken into account for this type of therapy is the count of monocytes and leukocytes as they increase, indicating, improvement of the bone marrow function. To be more informative we have to mention the existence of the management algorithm [11]. On the other side, Forghieri et al describe the therapeutic process of appendicectomy in phase one, at the time of diagnosis, showing off the safety and low complication risk, even in patients with autologous bone marrow transplantation during the aplastic crisis [12]. In our case report, which was a patient with gangrenous appendicitis in the context of neutropenia and immunosuppression, the scenario of extramedullary localization was blocked, based on the imagistic findings, as well as, the epidemiological data about the disease, even if the answer was given from the beginning. Concluding that appendicectomy is the only option in such cases, accompanied by the previously mentioned clinical image and imagistic findings. A special reference has to be made in the way of performing the appendiceal resection, nowadays, the preferred technique is the laparoscopic appendectomy, which offers a variety of advantages [23]. In our case, the laparoscopic appendectomy was initially attemped to be performed and finally converted to an open surgery. Even in patients with aplastic crisis and low levels of white blood cells, it is recommended immediate intervention via laparoscopic appendectomy.

## Conclusion

The surgeon must take into account the entire hematological conditions that may provoke acute appendicitis in order to be able to make the right decision for each patient individually. Especially, when talking about multiple myeloma, where the physician must know the changes in the epidemiological data, according to age, the co-existence of leucopenia and immunosuppression, the possibility of cecitis existence, which may be treated conservatively, and the possible extramedullary localization of the disease, which is extremely rare. The laparoscopic appendectomy constitutes the gold standard in the management of these conditions, even if it is performed immediately, in cases of aplastic crisis. To achieve the highest percentage of satisfying results in such cases, the multidisciplinary (radiologists, hematologists, general surgeons, and histopathologists) approach is considered the key to the way to success.

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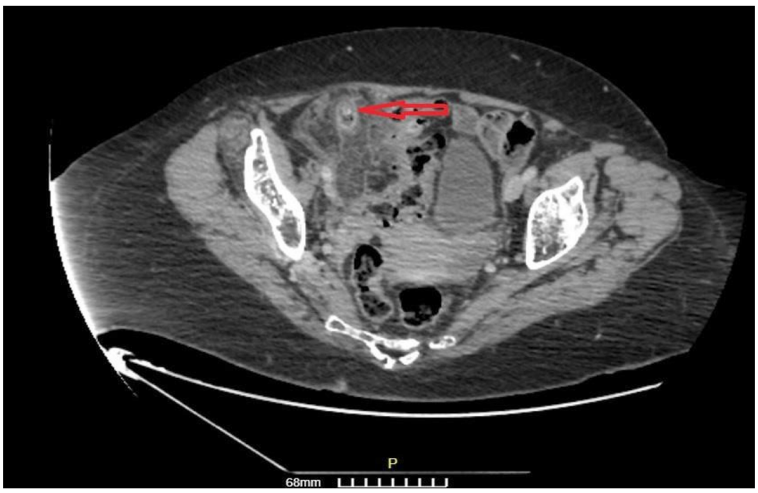
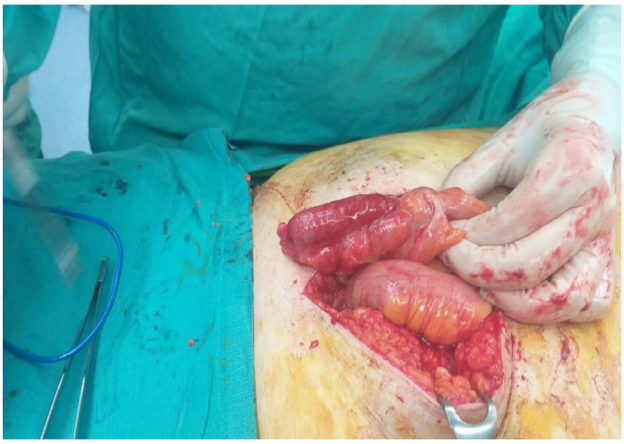
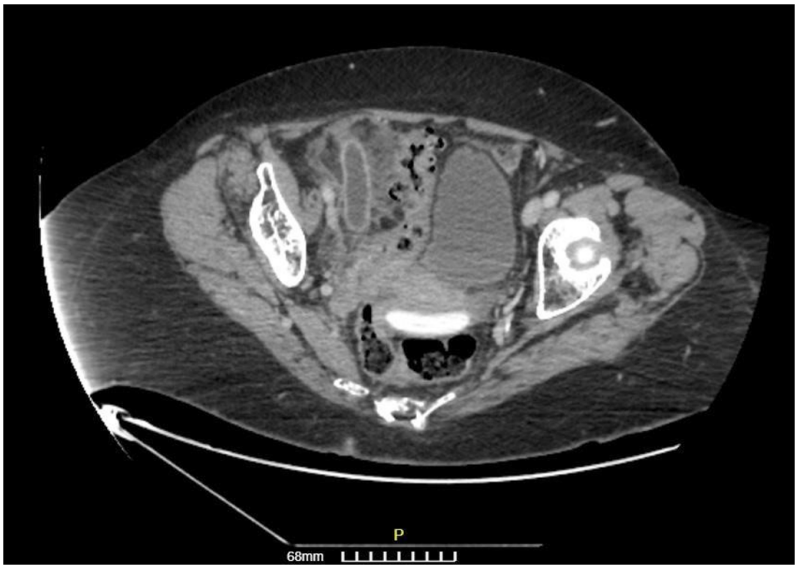
## Appendix




Intraluminal mass at the base of the appendix, suggestive a fecalith. D/d extramedullarylocation of the disease.



Enlarged appendix, with inflammation, wall thickening and concomitant mucosal damage.Adjacent to it is a normal part of small bowel.




Enlarged appendix, with wall thickening and accompanying mucosal damage. Dirty fatstranding of peripheral fat and epithelial pouch. Tape due to uterine prolapse.