Felty's Syndrome: A Qualitative Case Study

Wendy Woolston
Lynne M. Connelly

Felty's syndrome is a triad of rheumatoid arthritis, splenomegaly, and neutropenia. This rare disorder is difficult to diagnose and produces many complications. The purpose of this descriptive qualitative case study was to provide a comprehensive, context-bound understanding of one patient's struggle with the condition.

Felty's syndrome was identified in 1924 by Augustus Roi Felty when one of his patients presented with the same symptoms as five other patients in surrounding hospitals (Green & Fromke, 1966; Hume, Dagg, Fraser, & Goldberg, 1964). As a noted complication of longstanding rheumatoid arthritis (RA), Felty's syndrome is manifested by the triad of RA, splenomegaly, and neutropenia. It affects only 1%-3% of patients with RA, making this a rare, often unidentified condition (Owlia, Newman, & Akhtari, 2014). The affected person typically has comorbid splenomegaly and neutropenia, but he or she can be diagnosed with Felty's syndrome in the absence of splenomegaly (Keating, 2016; Owlia et al., 2014). The pathophysiology of the disease is not understood fully, but splenic sequestration and subsequent granulocyte destruction are believed to occur (Keating, 2016). Owlia and colleagues (2014) further noted, "…associated neutropenia is multifactorial including increased neutrophil sequestration secondary to splenomegaly…and failure of bone marrow to produce neutrophils" (p. 130).

Most patients diagnosed with Felty's syndrome are ages 50-70 and have had RA for more than 10 years (Keating, 2016; Rozin, Hoffman, Hayek, & Balbir-Gurman, 2013). Although Felty's syndrome is three times more common in women, men usually have earlier onset of symptoms. The disease is most common among Whites, and is rare in Blacks and children (Keating, 2016). A family history of RA increases a person's risk of developing Felty's syndrome (Xiao, Xiong, Long, Fan, & Lin, 2013). The condition often develops during a time when a patient is experiencing a remission of RA symptoms (Owlia et al., 2014; Rozin et al., 2013). It usually is not associated with juvenile idiopathic arthritis (JIA, formerly juvenile RA). Before 1998, only three girls with JIA had been diagnosed with Felty's syndrome; in two other cases, JIA persisted into adulthood and became Felty's syndrome (Balint & Balint, 2004; Xiao et al., 2013).

Pulmonary and skin infections are the most common complication of Felty's syndrome (Keating, 2016). Research supports the association of greater rates of infection with high incidence of mortality and increased risks of malignancy following diagnosis of Felty's syndrome (Balint & Balint, 2004; Xiao et al., 2013). The overall prognosis of Felty's syndrome is not favorable; mortality is higher because of the increased risk for serious infection (Owlia et al., 2014). The number of patients hospitalized with Felty's syndrome in the United States has declined over the past 20 years, possibly related to earlier and more aggressive treatment of RA; however, this hypothesis has not been supported with empirical data (Balint & Balint, 2004; Keating, 2016).

Treatment

The first line of treatment is the use of disease-modifying antirheumatic drugs such as methotrexate and hydroxychloroquine (Owlia et al., 2014). Owlia and colleagues initially found methotrexate dramatically increased the neutrophil values, but had varying success. In conjunction with splenectomy, the second line of treatment involves use of colony-stimulating factors (CSFs) to increase white blood cell (WBC) production. Although significant side effects often are encountered with CSFs (e.g., nausea, malaise, generalized pain, vasculitic rash), this treatment is effective at increasing neutrophil values in most patients (Owlia et al., 2014). Other treatment options available are parenteral gold, corticosteroids, intravenous immunoglobulins, plasmaphoresis, and prophylactic antibiotics (Balint & Balint, 2004; Xiao et al., 2013).

Wendy Woolston, MSN, RN, is Assistant Professor, Benedictine College, Atchison, KS.

Lynne M. Connelly, PhD, RN, is Associate Professor and Director of Nursing, Robert J. Dehaemers Endowed Chair, Benedictine College, Atchison, KS.

Purpose

The purpose of this case study was to develop a comprehensive, context-bound understanding of one patient's struggle with Felty's syndrome.

Literature Review

An online search was conducted of full-text, peer-reviewed scholarly journals on CINAHL, PubMed, Medline, and multiple Ovid sites to capture medical and nursing literature. With *Felty's syndrome* as the search term, 399 articles published between 2010 and October 2016 were located. The search then was modified to focus only on nursing; two articles about RA were identified (Nelson, 2011; Primdahl, Clausen, & Horslev-Petersen, 2013). Both articles mentioned Felty's syndrome as a complication but did not address it specifically. This illustrates the lack of nursing literature from the patient's point of view.

Sample Selection

This study used a purposive sample of one community-dwelling person with Felty's syndrome known to the principal investigator (PI). Initial contact was made with a member of her family to determine interest in participation, followed by telephone contact with the potential subject. The subject expressed an interest in sharing her story in hopes of helping others with Felty's syndrome.

Ethics

The study was approved by the Institutional Review Board at a private college in the midwestern United States. Investigators obtained informed consent at the beginning of the study and process consent was obtained verbally each time the participant was interviewed. The pseudonym *Carol* was used to identify the participant.

Methods and Design

A qualitative case study design was used to describe the participant's experience with Felty's syndrome. Case study research involves investigation and analysis of a single case to illustrate its uniqueness, particularity, and complexity (Hyett, Kenny, & Dickson-Swift, 2014). A case study approach was particularly appropriate for this study because of the rarity of the condition and the limited nursing literature. In addition, case studies are a practical approach to in-depth exploration of a patient's experience.

TABLE 1.
Semi-Structured Interview Questions

1.	Thinking back, prior to your diagnosis, how was your overall health?
2.	During the hospitalization in which you were diagnosed with Felty's syndrome, what were the health problems or issues you were experiencing?
3.	How did you feel when you received the diagnosis?
4.	How has Felty's syndrome affected your overall health and everyday life?
5.	As a nurse, I had never heard of Felty's syndrome before. What do you think nurses should know about caring for someone with this diagnosis?
6.	Who makes up your support system? How do they support you?
7.	Is there anything else that you would like to share about your experience?

Analysis

Data were collected through three interviews (over 150 minutes) with observation. The first interview was conducted at Carol's home, when observations were made of Carol's condition and surroundings. Her house was clean, well-organized, and free of clutter, but she was required to climb a flight of 22 stairs to get into her home. Lighting was low but adequate for reading. The temperature in the house was comfortable to the interviewer. Carol was clean and well-groomed, wearing sweatpants and a long t-shirt with a robe. In addition, she had a blanket across her lap. During the interview, she was sitting in a recliner in her living room with her feet elevated.

Semi-structured questions were used initially (see Table 1), but Carol was allowed to share her experiences with Felty's syndrome in her own way. When needed, probes were used to elicit details (e.g., "Tell me more about your experience with your family, healthcare providers, and hospitalizations"). After the researcher's initial analysis of data from the first interview, additional questions were developed to clarify the data. During the second interview, Carol was asked, "Who do you rely on for support?" "How did the doctors and nurses caring for you provide support?" "Describe how you have coped emotionally and physically with Felty's syndrome." After analysis of the first and second interviews and observations, a short third interview was conducted for member checking. Carol agreed her symptomology and feelings were captured correctly, and findings described her experiences.

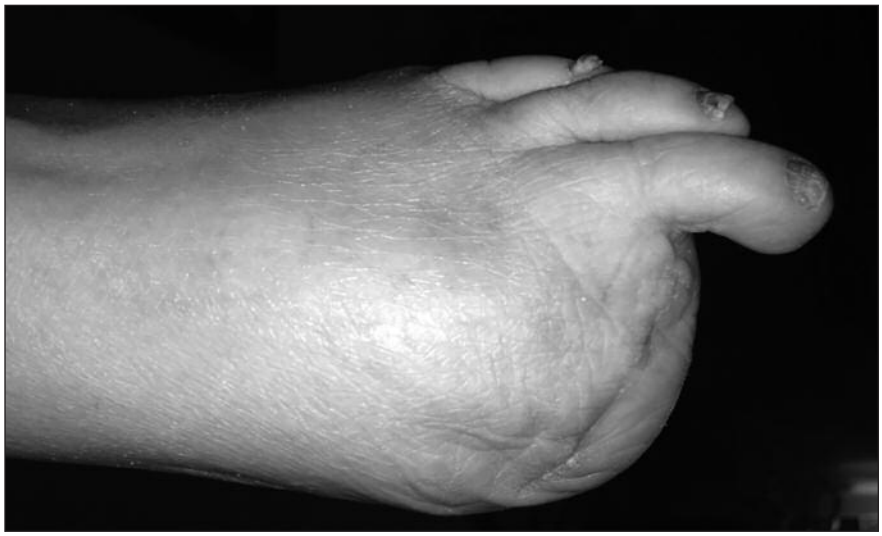
Interviews were audio-recorded and transcribed verbatim by the PI. All recordings and transcriptions were stored in a locked file cabinet in the PI's office. The first author read the text as a whole to get an overall sense of the data, and then re-read it and broke the data into meaning units for coding. Meaning units are the smallest pieces of data (e.g., groups of words) that contain useful information to address the research purpose (Bengtsson, 2016). All meaning units then were compared and contrasted, and coding was refined. The codes were reviewed and grouped into categories from which themes were developed.

Trustworthiness

Rigor in qualitative case study research is similar to other forms of qualitative research, including credibility, transferability, dependability, confirmability, and authenticity (Bengtsson, 2016; Cope, 2014; Guba

& Lincoln, 1994). In this study, credibility was enhanced by multiple interviews, observations, and member checking. Member checking with Carol consisted of reviewing themes and clarifying her description of the course of her medical condition. Process and methodological notes were written during the study to address dependability and confirmability. In addition, peer debriefing to review data analysis and discuss other issues was conducted periodically with an experienced qualitative researcher to minimize bias and ensure no relevant data were excluded from analysis and theme development. To enhance transferability and authenticity, authors described Carol's medical progress in detail from her perspective and included the interpretation of her experience with Felty's syndrome.

FIGURE 1.
Carol's Left Foot



First and second toe amputated, believed to be due to the combination of Felty's syndrome and diabetes.

Findings/Discussion

Patient's Medical Progression

At age 27, Carol awoke with severe pain in her hand. Although she tried to follow her usual routine of cleaning her house, the pain in her hand was so severe she thought she had a fracture. After a visit to her physician, she was diagnosed with RA. At that time, the doctor indicated she would probably be in a wheelchair before she was 36.

Carol was not in a wheelchair at age 36 but had fought a difficult battle. She was seeing an oncologist, who believed she had leukemia. Although she never had a confirmed diagnosis of leukemia, she had related symptoms: splenomegaly, neutropenia, and anemia. She had many respiratory infections that often were attributed to her smoking. For Carol, a simple cold would take 3-4 weeks to resolve. She had multiple wounds that would not heal. Something as simple as a paper cut would blister and take 6-7 weeks to heel. One particular wound started as a small puncture on her leg and did not heal for over 3 years. This wound was the size of a softball at its largest.

Because of the undiagnosed Felty's syndrome, she had multiple invasive treatments without improvement. She was selected to receive an Apple Graft, a rare skin graft technique that uses the foreskin from infant circumcisions to cover the wound. Unfortunately, her body rejected the graft. Even when infections were present, her WBC count was low. The lowest count she could recall was 900 cells/microliter (normal 4,500-10,000 cells/microliter) (Kee, 2014).

At age 46, Carol was diagnosed with Felty's syndrome. She was relieved to have a diagnosis but also afraid because she and the nurses caring for her had never heard of this rare condition. Carol reported, "…I became even more scared when I looked it up. What is even harder is that there is no one around here with Felty's syndrome I can talk to." Three years after the diagnosis, three physicians agreed the best treatment for Carol was a splenectomy. Leaders of the hospital where she was treated would not allow an elective splenectomy without three physicians agreeing this was the best treatment.

Following the splenectomy, her WBC count increased to 6,000-7,000 cells/microliter. She was told by her physicians her spleen weighed about 5 pounds (normal 3.2-3.7 pounds). The wound on her leg significantly decreased in size during the first postoperative week and was healing by the second week. Age 55 at the time of the interviews for this case study, Carol continued to have residual effects of Felty's syndrome. She had numerous methicillin-resistant *Staphylococcus aureus* infections, the first starting from an intravenous site in her arm. She had two toes amputated, and the healed leg ulcer still opens if her leg swells (see Figures 1 & 2). Her fingernails and toenails were hyperpigmented (see Figure 3). She also received a definitive diagnosis of leukemia common in Felty's syndrome (Liu & Loughran, 2011; Shiel, 2016). See Table 2 for her medical and surgical diagnoses.

Carol lived in a rural area with her husband. She had eight siblings with five still alive; none have RA. Her maternal grandmother had RA. Carol had four grown children (three daughters, one son) and 12 grandchildren who lived in the area. One daughter had been diagnosed with RA shortly before the interviews for this case study. Carol lived in a mobile home with a basement. Because the back entrance to her house has many stairs, her husband would drive her to the front door so

FIGURE 2.
Healed Ulcer on Carol's Lower Right Leg

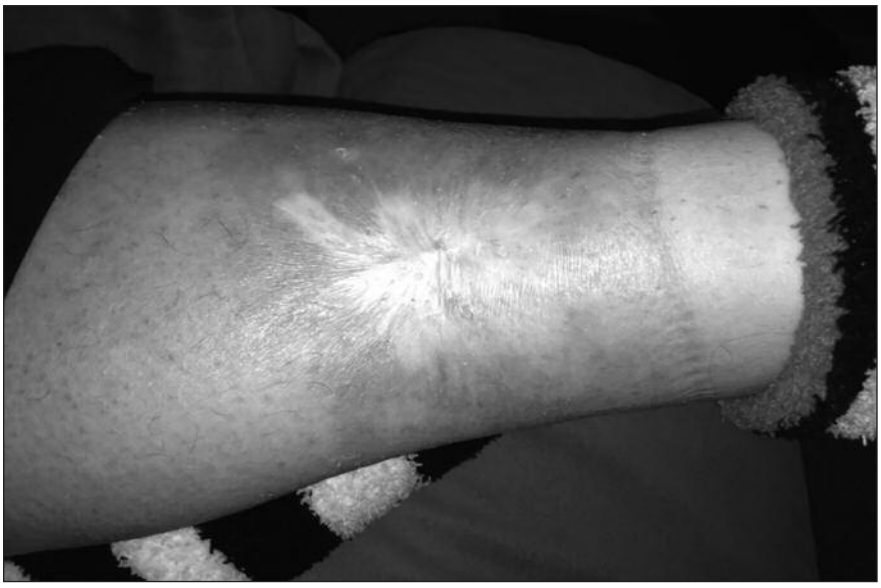
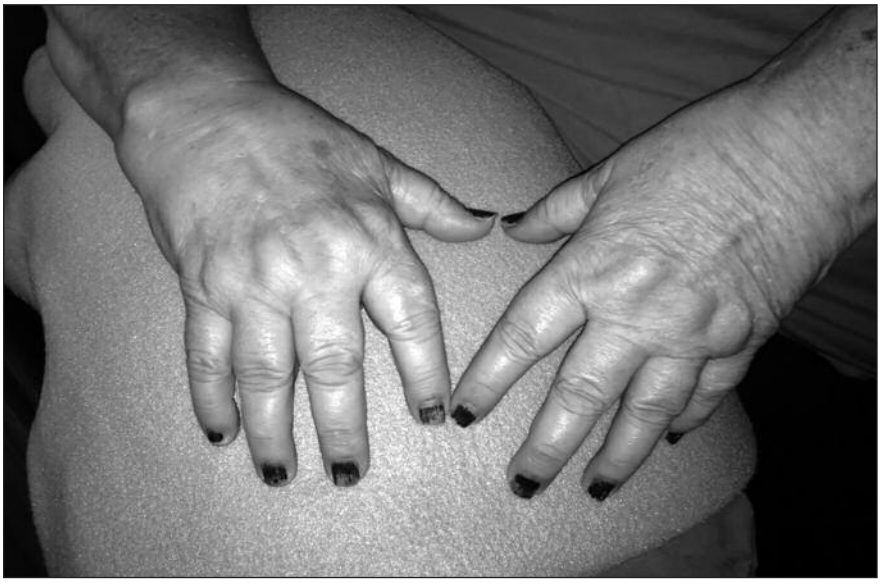


FIGURE 3.
Carol's Hands



Nail polish is worn to cover up the hyperpigmentation of her nails.

she only had a few stairs to climb. She used a computer chair with wheels to move around her house.

Carol's experience with this condition affected nearly all aspects of her life. The overarching theme for this case study was *Felty's messes with everything*. Other major themes that emerged from the data included *managing pain that never goes away*, and a *lack of support and understanding and feeling scared and lonely*.

Managing Pain that Never Goes Away

Carol described unrelenting pain that was difficult to manage. She rated her pain intensity as 8-10 (on 0-10 intensity scale) all the time. "My pain is more severe than the pain of childbirth and it never goes away," she commented. This affected her ability to do the simplest activities, such as housework, cooking, and washing dishes. At one point, she asked to have the fentanyl transdermal patch (Duragesic®) prescription discontinued. She experienced difficulty taking the drug because she would forget to replace the patch or forget to remove the old patch. She also had panic-type attacks, calling her children at all hours. She decided to go to a drug abuse clinic and was given methadone to help her discontinue the patch. She wondered, "I cannot believe a doctor gave me a medication that could be so addictive."

She described attending a pain meeting of approximately 4 hours.

> By the time I left, I was late on my next pain pill and in severe pain. We have to sign papers stat-

TABLE 2.
Medical and Surgical History

Felty's syndrome
Splenomegaly
Back pain
Chronic obstructive pulmonary disease
Diabetes mellitus type 2
Diabetic neuropathy
Fibromyalgia
Gastroesophageal reflux disease
Hypertension
Inflammatory neuropathy
Leukocytosis
Obesity
Osteomyelitis
Pancytopenia
Plantar fasciitis
Seizures
Rheumatoid arthritis
History of pneumonia
Methicillin-resistant *Staphylococcus aureus*
Carpel tunnel syndrome
Bilateral knee arthroplasty
Amputation of toes
Splenectomy

> ing we won't sell or give our drugs away. Why would I sell it? I need it. I won't overuse it. I take three pills a day, no more or less.

Dealing with unrelenting pain was frustrating for Carol and led her to be angry at the disease, sometimes at God, but she generally managed to stay positive. "Every day I wonder and sometimes pray I do not wake up. Then I remember my family. I have things to look forward to." In addition, she stated, "There are people a lot worse than me. I just read in the paper about a woman who is paralyzed from the neck down." Her WBC count affected the pain. When the count was low, she had less pain. When the value was as high as 17,000 cells/microliter, she experienced a great deal of pain. Her goal was a WBC count of 11,000 cells/microliter, with the lowest desired at 9,000 cells/microliter.

A Lack of Support and Understanding and Feeling Scared and Lonely

Carol described her relief when she finally received the diagnosis of Felty's syndrome. She also was scared because she had never heard of it, and became even more afraid when she researched the condition. What was particularly hard for her was having no other person with Felty's syndrome in her life for conversation. "My doctors even tell me they do not know anyone with it."

The lack of support from other sufferers made the experience lonely for Carol. She perceived her family did not understand her condition and avoided her at times. She particularly thought her husband did not understand her experience. She stated, "I don't get much support. My family doesn't want to hear about it." Carol believed she has missed a great deal, particularly with her children and grandchildren. "I am sure my grandkids say, 'Why did grandma not do more?'" Carol spent most of her time at home and found it difficult to move around the house. She worried others would think she was lazy. She described feeling isolated and alone with this condition. She also expressed fear for her daughter, who recently was diagnosed with RA: "I am afraid my daughter will get it (Felty's syndrome)."

Overall Theme: Felty's Syndrome Messes with Everything

Unfortunately, the physical effects were only part of Carol's suffering. Emotionally, the day-to-day effects of RA and Felty's syndrome could seem unbearable. As Carol noted,

> Felty's syndrome messes with everything. Sometimes I get so frustrated and angry. I often feel left out because I cannot do as much. I cannot go shopping with the kids. I have really missed out on a lot of milestones in my kids' and grandkids' lives. I am sure my grandkids wonder why I did not do more with them. This causes me to get depressed and it gets harder every day. Some people just think I am lazy. I love to do housework, but I cannot stand at the sink long enough to do the dishes. Just to move around the house, I use my computer chair.

In the interview, Carol indicated one of her greatest desires was to have more family support. Although one of her daughters was very supportive and helped her a great deal, she believed her two other daughters and son did not want to spend time with her. Carol said, "Just a phone call every now and then would be enough." She said, "… sometimes your family forgets that even though you are not in the hospital you are still not well." She further explained, "You hate to intrude on your kids at night because they are busy with their families."

This study added to the literature about Felty's syndrome by presenting the patient's experience in struggling with her illness. The themes in this study demonstrated important concerns for Carol. In addition, the interviews appeared to be therapeutic for her and she seemed to want to share her experience in a way that could help others.

Nursing Implications

Education, infection control, and pain management are the most important nursing interventions for persons with Felty's syndrome (Nelson, 2011; Shiel, 2016). Patients are at high risk for infection, slower wound healing, and surgical incision complications (e.g., bleeding, dehiscence). They need to be educated about the warning signs of delayed wound healing, infection, surgical incision bleeding and dehiscence, and increased WBC count (Nelson, 2011; Shiel, 2016). The nurse should ensure patients have access to medical care if infection occurs. Patients additionally must understand the disease course, realizing symptoms will be lifelong and continue to worsen over time. Family education is also important. Family support can help patients face the day-to-day struggles. Families need to be educated about the illness, pain intensity, and physical disability their loved ones will experience. Online support groups can provide an opportunity for patients and family members to share personal struggles and accomplishments with others with a similar experience. The pain of RA is often intense, and can be unbearable when coupled with Felty's syndrome. The nurse should ensure patients receive analgesia as scheduled and do not exhaust their prescriptions.

Limitations

This single case study related to one woman's struggle with Felty's syndrome cannot be generalized beyond the context of this case. In addition, the participant had difficulty remembering specific facts and dates, and had multiple health conditions that could have contributed to some inaccurate data. Further research is needed to support the findings of this case study, but may be difficult due to the rarity of this disease process.

continued on page 118

continued from page 109

Conclusion

Nurses should have basic knowledge of Felty's syndrome. Because of the rarity of the disease, it may be overlooked or misdiagnosed as other conditions (e.g., leukemia, systemic lupus erythematosus) (Liu & Loughran, 2011; Shiel, 2016). Nurses should recognize patients with RA who also have nonhealing leg ulcers and low WBC counts may be demonstrating early symptoms of Felty's syndrome (Owlia et al., 2014). MSN

REFERENCES

Balint, G.P., & Balint, P.V. (2004). Felty's syndrome. *Best practice & research clinical rheumatology*, *18*(5), 631-645.

Bengtsson, M. (2016). How to plan and perform a qualitative study using content analysis. *NursingPlus Open, 2,* 8-14.

Cope, D.G. (2014). Methods and meanings: Credibility and trustworthiness of qualitative research. *Oncology Nursing Forum, 41*(1), 89-91.

Green, R., & Fromke, V.L. (1966). Splenectomy in Felty's syndrome. *Annals of Internal Medicine*, *64*(6), 1265-1270.

Guba, E.G., & Lincoln, Y.S. (1994). Competing paradigms in qualitative research. In N.K. Denzin & Y.S. Lincoln (Eds.), *Handbook of qualitative research* (pp. 105-117). London, England: Sage.

Hume, R., Dagg, J.H., Fraser, T.N., & Goldberg, A. (1964). Anemia of Felty's syndrome. *Annals of Rheumatic Diseases*, *23*, 267-271.

Hyett, N., Kenny, A., & Dickson-Swift, V. (2014). Methodology or method? A critical review of qualitative case study reports. *International Journal of Qualitative Studies in Health and Well-Being, 9.* doi:10.3402/ghw.v9.23606

Keating, R.M. (2016). *Felty syndrome.* Retrieved from http://emedicine.medscape.com/article/329734-overview

Kee, J.L. (2014). *Laboratory and diagnostic tests with nursing implications* (9th ed.). Upper Saddle River, NJ: Pearson.

Liu, X., & Loughran, T.P. Jr. (2011). The spectrum of large granular lymphocyte leukemia and Felty's syndrome. *Current Opinion in Hematology, 18*(4), 254-259.

Nelson, D.E. (2011). Perioperative care of the patient with rheumatoid arthritis. *AORN Journal, 94*(3), 290-300.

Owlia, M.B., Newman, K., & Akhtari, M. (2014). Felty's syndrome, insights and updates. *The Open Rheumatology Journal, 8,* 129-136.

Primdahl, J., Clausen, J., & Horslev-Petersen, K. (2013). Results from systematic screening for cardiovascular risk in outpatient with rheumatoid arthritis in accordance with EULAR recommendations. *Annals of Rheumatic Diseases, 72*(11), 1771-1776.

Rozin, A.P., Hoffman, R., Hayek, T., & Balbir-Gurman, A. (2013). Felty's syndrome without rheumatoid arthritis? *Clinical Rheumatology, 32*(5), 701-704. doi:10.1007/s10067-012-2157-3

Shiel, W.C. (2016). *Felty's syndrome.* Retrieved from http://www.medicinenet.com/feltys_syndrome/article.htm

Xiao, R., Xiong, M., Long, Z., Fan, R., & Lin, D. (2013). Diagnosis of Felty's syndrome, distinguished from hematological neoplasm: A case report. *Oncology Letters, 7*(3), 713-716.