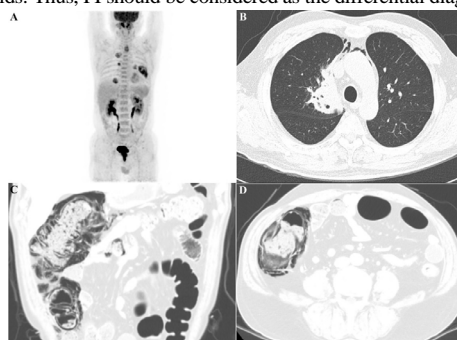


Granulomatosis with Polyangiitis complicated by Pneumatosis intestinalis and Pneumomediastinum

¹부산대학교병원, 내과, ²부산대학교병원, 호흡기알레르기내과, ³부산대학교병원, 류마티스내과

*김아란¹, 엄중섭², 고정희³

Granulomatosis with polyangiitis (GPA, formerly Wegener's Granulomatosis) is a systemic small vessel vasculitis, characterized by necrotizing granulomatous inflammation and is often associated with anti-neutrophil cytoplasmic antibodies (ANCA). The main clinical manifestations involve the upper and/or lower respiratory tract and kidneys. A 66-year-old man, who was diagnosed with GPA involving lung, left orbit and nasopharynx 4 months ago, was admitted for the fourth cyclophosphamide therapy session (Figure 1A). He had presented blood tinged sputum for 2 weeks. The laboratory test showed elevated proteinase-3 ANCA titer (61.3 U/mL) and C-reactive protein (2.32 mg/dL) compared to previous study. The chest computed tomography (CT) scan revealed that increased size of a cavitary lung mass, nodular consolidation, new onset of pneumomediastinum and pneumoperitoneum (Figure 1B). Additional abdominal CT scan showed pneumatosis intestinalis (PI) of ascending colon (Figure 1C and 1D). The patient was treated conservatively with bowel rest, intravenous antibiotics and oxygen therapy, as the patient was asymptomatic. PI is defined as the presence of gas within the wall of the gastrointestinal (GI) tract. A vasculitis-based pathogenesis within the intestinal territory, a GI tract involvement resulting from a vascular injury, or treatment with glucocorticoids could be associated with development of PI in the various rheumatic diseases. Although many patients with rheumatic diseases who develop PI respond to conservative therapy, treatment decisions must be based on the patient's underlying condition. Herein, we describe our experience with one such case of PI in a GPA patient who was previously treated with cyclophosphamide and glucocorticoids. PI is a rare complication of rheumatic diseases, particularly ANCA-associated vasculitis. However, patients with ANCA-associated vasculitis have multiple risk factors for developing PI, including GI vasculopathy and treatment with glucocorticoids. Thus, PI should be considered as the differential diagnosis.



Tenosynovial giant cell tumor In systemic lupus erythematosus

¹아주대학교 병원 내과부, ²아주대학교 병원 류마티스 교실

*송용희¹, 김현아^{1,2}, 서창희^{1,2}, 정주양^{1,2}

Tenosynovial giant cell tumors are a group of generally benign intra-articular and soft tissue tumors with common histologic features. These tumors cause the affected synovium, bursae, or tendon sheaths to thicken and overgrow. Symptoms can include pain, swelling, and limitation of movement of the joint. If untreated or if the tumor continually recurs, these tumors can result in damage and degeneration of the affected joint and surrounding tissues or structures. we present a case report of tenosynovial giant cell tumors in systemic lupus erythematosus patient. In May 2015, a 44 years old woman who had not been diagnosed with any kind of disease before visited the Rheumatology clinical office for multiple joints pain and swelling that started a month ago. Through the workup, she was diagnosed with systemic lupus erythematosus due to inflammatory arthritis (tender joint count 4, swollen joint count 4), leukopenia, ANA+, Ro+, APA+, oral ulcer). She started the therapy with hydroxychloroquine, glucocorticoids, and NSAID. Her arthritis got improved after addition of tacrolimus. In March 2017, she complained that her arthritis was getting worse and a mass like lesion appeared on her Lt foot dorsum. On articular ultrasound, the mass was confirmed with high signals of power Dopplers (PDs) and blood flow. (Fig. 1) Lt foot MRI was taken and it showed us About 3.4 * 2.0 cm sized lobulated contour, dumbbell shape mass between Lt 3rd web space. (Fig. 2) This mass was widening of 3rd web space and abutting extensor digitorum tendon, 3rd and 4th. The mass excision was done, and the specimen consists of a fragment of greyish brown firm tissue, measuring 3.9x2.7x1.7cm. (Fig. 3) Historical result was Tenosynovial giant cell tumor, localized type. This is a rare case about a combined Tenosynovial giant cell tumor in patients with SLE. Clinical presentation of Tenosynovial giant cell tumor is nonspecific, and it should be considered in the absence of evidence for any other synovial pathology. Currently, surgery remains the treatment of choice for patients with Tenosynovial giant cell tumors and it is maximal resection of pathological tissue.



<Fig. 1> The ultrasound of Lt foot dorsum

<Fig.2> Left foot MRI showed 3.4 * 2.0 cm sized lobulated contour mass between Lt 3rd web space.

<Fig. 3> The specimen