



Syndrome of Remitting Seronegative Symmetrical Synovitis with Pitting Edema (RS3PE): A Case Report and Review of the Literature

Semra Akturk

Adiyaman University Education and Research Hospital, Clinic of Physical Therapy and Rehabilitation, Adiyaman, Turkey

Abstract

Remitting Seronegative Symmetrical Synovitis with Pitting Edema (RS3PE) is a rare clinical entity that is easily missed due to lack of knowledge. Syndrome is characterized by sudden onset of edema with swelling on the dorsum of the hands and by synovitis. Serological tests are negative and radiographic joint destruction does not occur. Etiology of RS3PE syndrome is not known. It can occur as an idiopathic phenomenon, but also is in association with rheumatic diseases. In addition, several reports have described RS3PE as a paraneoplastic syndrome can occur concomitantly with both hematological and solid malignancies. Because of this clinical association, it is not very easy to diagnose this syndrome. We discuss a case of RS3PE where the patient presented with acute onset polyarthritis and pitting edema of the extremities without an underlying systemic cause. Patient showed dramatic response with low dose steroid therapy.

Keywords: RS3PE, seronegative, symmetric arthritis

(Rec.Date: June 10, 2015 Accept Date: June 11, 2015)

Corresponding Author: Semra Akturk, Adiyaman University Education and Research Hospital Physical Therapy and Rehabilitation Clinic, Adiyaman, Turkey

E-mail: semrakayakturk@hotmail.com.tr

Introduction

Remitting seronegative symmetrical synovitis with pitting edema (RS3PE) is described as a syndrome suddenly appearing edema in dorsal side of hands or feet and flexor tendinitis of fingers affecting mostly wrist or ankle joints in a symmetrical fashion and it has a good prognosis. This syndrome generally affects elderly population and has a male predominance with a male/female ratio of 4. It has been defined as one of subgroups of seronegative rheumatoid arthritis (RA), and its etiology has not been understood completely. Absence of serum rheumatoid factor (RF) and articular erosion on direct radiography are typical findings. The most important feature is a good response to steroid therapy without any joint sequelae [1,2].

Because of its rarity, this syndrome is not usually considered in the differential diagnosis, and patients may be exposed to various unnecessary clinical tests and may subsequently receive different treatments. An important point which should be paid account is that RS3PE syndrome may appear as a neoplastic fashion in various hematological and solid malignancies or together with rheumatic diseases [3,4].

Case

A 60-year-old male patient was presented with with complaints of pain and swelling in both wrists and dorsal parts of hands and restriction of motion and swelling in ankles for about two weeks. The patient stated that he had morning stiffness in fingers that lasts more than one hour.

The patient had hypertension that is under control with amlodipine 10 mg once-daily dose of 10 mg for about 8 years. The patient went to an internal medicine specialist after the beginning of the complaints in hands and feet, who said that amlodipin molecule may cause some swellings in hands and feet and the drug should be changed. Because any improvement in the status of the patient did not occur after the change of drug and going several other clinics, the patient was referred to our clinic. On examination, there was tenderness and increased temperature in each wrist, metacarpophalangeal and proximal interphalangeal joints and ankles. Marked diffuse pitting edema was present in the dorsum of each foot (+2) especially being more prominent in wrists (+3) (Figures 1a and 1b).



Figures: 1a: edema of the dorsum of the hand, 1b: ankle edema

The remainder of the physical examination was otherwise normal. Laboratory tests were performed to narrow the differential diagnosis while taking into account RS3PE. Complete blood count, routine biochemical tests and urinalysis was found within normal ranges. Serum parathormone was 188 pg/mL (normal range 19.8-74.9 pg/mL), sedimentation rate 48 mm/h, RF 4.2 IU/mL (normal value <20 IU/mL), C-reactive protein 1.1 mg/dL (normal range 0.01-0.82 mg/dL). Thyroid, renal and liver function tests were within normal limits. Computerized tomography scan of thoracoabdominal region for the search of any malignancy was reported as normal. On anterior-posterior view of hand-wrist plain graphy, there were osteophyte formation, periarticular osteopenia and narrowing of bilateral proximal and distal interphalangeal joint spaces.

In the light of these findings, the patient was considered as RS3PE syndrome. Once-daily oral dose of 10 mg prednisolon treatment was started and the patient was called for a control visit. Within several days, marked improvement in clinical status of the patient was observed after the beginning of the treatment.

Discussion

Diagnostic criteria for RS3PE syndrome was firstly defined by Olive et al as being over 50 years old, pitting edema in the dorsum of both hands, sudden onset of polyarthrititis and RF negativity [5].

As in our case, diagnosis can easily be made with these criteria. In addition, dramatic response to low-dose steroid treatment (10-20 mg) and healing without any sequel and remaining in remission for a long time also support the diagnosis.

After the definition by McCarty in 1985, more than 150 case reports have been cited in the literature. When all those cases are examined, it can be seen that making diagnosis of RS3PE with criteria cited above contains some difficulties;

It is unclear whether this syndrome is a specific disease or an initial form of rheumatic disease or a disease associated with other rheumatic diseases [6]. It will not be easy to narrow the differential diagnosis in this circumstances. In particular, when some patients who does not enter remission despite the low dose steroid therapy, one of RA, Sjogren's syndrome, systemic lupus erythematosus and vasculitic syndromes has been shown to develop over the course of the disease [7,8].

RS3P syndrome may occur as a paraneoplastic syndrome of various malignancies. This condition may develop before, simultaneously or after the development of malignancy. In patients diagnosed with this syndrome, paraneoplastic syndromes should be ruled out and patients should be monitored for a long time, even for life-long if necessary [3,9].

When considering the healthy subjects in the same age group of those with RS3P, concomitant presence of various systemic symptoms and coexistence of many systemic and chronic diseases in the elderly population, the diagnosis of RS3P may be challenging.

Although the disease is a symmetric fashion, there are several case reports associated with unilateral joint involvement of RS3PE syndrome recently [10].

Although these diseases are frequently observed in men and people over age 50, there are considerable number of case reports also in earlier ages and females [11,12].

Laboratory tests frequently show RF negativity, mild and moderate acute phase response, anti-nuclear antibodies (ANA) negativity and presence of HLA-B7, B22 and B27 tissue antigenin some patients [13,14]. Because of these laboratory findings may occur in a variety of situations, it is not diagnostic for this syndrome.

Although there exist absence of erosion in direct X-ray graphic and nonspecific tenosynovitis on ultrasound and MRI, these findings are not diagnostic.

In conclusion, RS3Pe syndrome is a disease characterized by acute onset polyarthritis, edema of dorsum of hands and feet, RF negativity and absence of erosion on X-ray graphics, good response to low dose corticosteroid treatment and remaining in remission for a long time. The association of this syndrome with a paraneoplastic syndrome and rheumatic diseases and also lack of clear diagnostic criteria may cause difficulties in making differential diagnosis of this syndrome. For these reasons, RS3PE syndrome should not be overlooked in differential diagnosis of patients demonstrating the characteristics listed above.

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