

A Condition Confused with Complex Regional Pain Syndrome in Hemiplegia: Simultaneous Occurance of Knee Osteonecrosis and Monoarthritis

Hemiplejide Kompleks Bölgesel Ağrı Sendromu ile Karışabilen Bir Durum: Eş Zamanlı Görülen Diz Osteonekrozu ve Monoartrit

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ABSTRACT Complex regional pain syndrome (CRPS) type 1 is a painful condition commonly seen in hemiplegia and affects negatively the rehabilitation process. We report a case of 42-year-old male with right sided hemiplegia who developed acute inflammatory monoarthritis at the right knee joint during the stroke rehabilitation program. The patient also had clinically silent knee osteonecrosis (ON) that noticed after the clinical and radiological examination period for the inflammatory monoarthritis. The cause of the monoarthritis was investigated but it remained indeterminate. The coexistence of these two conditions led to a diagnostic confusion with complex regional pain syndrome type 1 (CRPS1). In this case report we aimed to stimulate further thought in the field of misdiagnosing CRPS1. We also discussed whether ON and inflammatory monoarthritis were related or not. We highlighted the importance of distinguishing CRPS and ON especially in stroke patients. A high index of suspicion could avoid diagnostic delay.

Keywords: Osteonecrosis; complex regional pain syndrome; hemiplegia; arthritis; rehabilitation

ÖZET Kompleks bölgesel ağrı sendromu (KBAS) tip 1 hemiplejide yaygın olarak görülen ve rehabilitasyon sürecini olumsuz etkileyen ağrılı bir durumdur. Bu yazıda, 42 yaşında sağ hemipleji tanılı, inme rehabilitasyon programı sırasında sağ dizinde akut inflamatuvar monoartrit gelişen bir erkek hasta sunuldu. İnflamatuvar monoartrite yönelik yapılan klinik ve radyolojik incelemeler sırasında hastada klinik olarak sessiz seyreden bilateral diz osteonekrozu olduğu fark edildi. İnflamatuvar monoartrite yol açan spesifik bir romatolojik hastalık tespit edilemedi. Osteonekroz ve monoartritin aynı anda bulunması başlangıçta hastanın KBAS olarak değerlendirilmesine yol açmıştır. Bu vakayı sunmadaki amacımız hemiplejide KBAS'la karışabilecek bir durumu ortaya koyarak, yanlış tanı konulmasını önlemede farkındalık yaratmaktır. Ayrıca osteonekroz ve inflamatuvar monoartrit birlikteliği bu iki durumun birbirleriyle ilişkili olup olmadığı açısından araştırma yapılması gerekliliğini doğurmuştur. Bu vaka sunumunda özellikle inme hastalarında KBAS tip 1 ve osteonekroz tanı ayırımının önemi ortaya çıkmaktadır. Bu iki hastalığın ayırıcı tanısına yönelik hassasiyet gösterilmesi tanı gecikmesini önleyecektir.

Anahtar Kelimeler: Osteonekroz; kompleks bölgesel ağrı sendromu; hemipleji; artrit; rehabilitasyon

Osteonecrosis (ON) is a progressive disease that often causes subchondral collapse and disability at the affected joint. Pain and reduced range of motion are usually revealed in symptomatic patients. ON of the knee is divided into three categories: 1. Spontaneous ON of the knee, 2. Secondary ON and 3. Post-arthroscopic ON. Secondary ON is associated with systemic steroid therapy, systemic lupus erythematosus, smoking, haemoglobinopathies, dyslipidemia and alcohol abuse.^{1,2} Complex

regional pain syndrome (CRPS) is another cause of knee pain and should not be ignored in the differential diagnosis of ON. CRPS is characterized by sensory-motor disturbances, discoloration of the skin, changes in temperature and sudomotor activity, vasomotor edema and trophic findings that are disproportionate to the initiating event.³ CRPS is divided into two types based on the presence of nerve lesion following an injury. CRPS type 1 (CRPSt1) may develop after stroke and affects mostly the upper extremity. The diagnosis is clinical and several sets of diagnostic criteria were developed including Budapest Criteria which is a widely used standardized criteria for clinical and research diagnosis. In this case report we present a stroke patient with simultaneous secondary knee ON and inflammatory monoarthritis that led to diagnostic confusion with CRPSt1. We aimed to stimulate further thought in the field of misdiagnosing CRPSt1.

CASE REPORT

A 42 year-old male was referred to our physical medicine and rehabilitation clinic with the diagnosis of right sided hemiplegia for seven weeks. The baseline Brunnstrom Motor Recovery Stages (BMRS) were 1 for hemiplegic upper and lower extremities and hand. Sensory examination revealed hypoesthesia at the affected side. A comprehensive rehabilitation program, including range of motion, progressive resistive exercises, balance and coordi-

nation training and neurophysiological approaches were planned. At the third week of the admission, an improvement was observed on the lower extremity's motor functions (BMRS 2) that the patient was started to verticalization and walking exercises in the parallel bars. At the first week of the verticalization; warmth, mild swelling and moderate pain without neuropathic features occurred at the right knee. The laboratory findings revealed high levels of sedimentation rate and C-reactive protein (72 mm/h and 8.9 mg/dl respectively) and leukocyte count ($10\,000/\text{mm}^3$). Bilateral multifocal osteopenic lesions at distal femur and proximal tibia were determined on the anteroposterior and lateral plain radiographs of the knees (Figure 1). Joint rest, compression, elevation, cryotherapy, transcutaneous electrical nerve stimulation and nonsteroidal anti-inflammatory drugs were used to reduce pain and inflammation. The initial diagnosis was considered as CRPSt1 of the knee according to the radiographic findings which suggested mottled osteoporosis and signs of inflammation in the hemiplegic side. But we needed to confirm the diagnosis because of the insufficient clinical features especially the non-neuropathic character of the pain. For this reason 3-phase bone scan (TPBS) was obtained. TPBS revealed increased both blood flow and blood pool at early phase and increased radiopharmaceutical uptake at the late phase bilaterally, compatible with CRPS. In the following days increase in inflammation was observed



FIGURE 1: Anteroposterior and lateral plain radiographs of bilateral knee joints demonstrated bilateral multifocal osteopenic lesions at distal femur, proximal tibia and fibula.

and aspiration of the knee joint was carried out. Synovial fluid analysis demonstrated inflammatory properties in microscopic and biochemical investigation (white cell count 32 000/mm³ (80% polymorphs), glucose: 70,5 mg/dl, protein: 4,7 g/dl). After the exclusion of septic arthritis, intra-articular corticosteroid injection was performed. The patient was assessed for possible causes of inflammatory acute monoarthritis including crystal-induced arthritis, rheumatoid arthritis, seronegative spondyloarthritis and connective tissue diseases but the findings didn't support these diseases. In the following days reduction in the signs of inflammation were observed and the patient restarted to ambulation training. We considered that the inflammatory monoarthritis mimicked the symptoms of CRPSt1 and we needed further investigation to find out the underlying reason of bilateral lytic lesions in the knees. Contrast-enhanced magnetic resonance imaging (MRI) was performed and revealed bilateral intramedullary lesions in distal femur, proximal tibia and fibula consistent with bilateral ON of the knees (Figure 2). It was also distinguished from the infiltrative diseases of the bone marrow by bone marrow biopsy and protein electrophoresis. By the light of these findings the patient was diagnosed as bilateral ON of the knee accompanied by unilateral inflammatory monoarthritis. Conservative treatment and outpatient follow-up were recommended by the orthopedic surgeon. At the end of the ten week rehabilitation program, the patient discharged at the level of standing and taking a few steps in the parallel bars.

Patient's written informed consent was obtained.

DISCUSSION

In this case report we presented a stroke patient with simultaneous secondary knee ON and inflammatory monoarthritis of the knee. The clinical and radiological examination period revealed underlying clinically silent ON of the knee. According to the coexistence of signs of inflammation and the plain radiographic findings, we considered CRPSt1, a common problem in stroke patients, as the initial diagnosis but the diagnostic criterias were not fully met.⁴

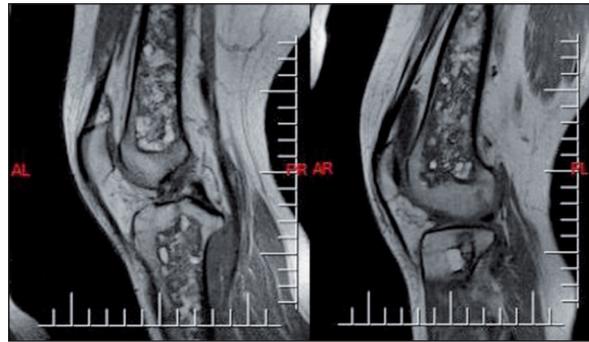


FIGURE 2: Sagittal T1 sequence images demonstrated iso-hyperintense intramedullary lesions in bilateral distal femur, proximal tibia and fibula.

Generally TPBS is performed in clinically doubtful CRPS which typically shows increased flow and pool activity and radiotracer uptake in the affected bones due to the increased bone turnover.⁵ Usually TPBS is also performed in diagnosing ON that various manifestations could be seen according to the disease stage. At early ON, a decrease in activity is observed in the necrotic area of the bone and the next period an increase in activity is developed due to the revascularization, increased both blood flow and osteoblastic activity.⁶ This variability may cause a difficulty in distinguishing the two diseases. In this patient because of the insufficient clinical features for diagnosis of CRPS, MRI was needed for further investigation.

MRI is proposed to be the most sensitive and specific imaging modality in the detection of osteonecrotic lesions.⁷ Mitchell et al. has developed a classification system for ON, according to the signal characteristics within the center of the osteonecrotic lesion.⁸ In this case report, the MRI findings of lesions were compatible with class B signal intensity (high intensity in T1 and T2 sequences). MRI may also be helpful to distinguish osteonecrosis from other bone disorders.⁹

Another compelling condition was the coexistence of ON and inflammatory monoarthritis that caused a question whether they were related or not. In the literature it is known that in ON, acute synovitis episodes could develop but non-inflammatory properties are proposed.¹⁰ In the light of this information we thought that a concomitant inflammatory rheumatic disease could be the rea-

son of the inflammatory monoarthritis. We investigated the cause of the monoarthritis but it remained indeterminate. In the literature related to this issue, only Rabquer et al. have investigated synovial tissue in patients with femoral head ON.¹¹ They suggested that the synovial fluid analyses of a large population of the patients revealed inflammatory without having coexistence of inflammatory disease. But in this case report, the patient's synovial fluid analysis revealed neutrophil dominance in leukocyte count incompatible with the mentioned study. According to these findings, we thought that the inflammatory monoarthritis was unrelated with ON. Further studies investigating

the synovial inflammation in osteonecrosis are needed.

CONCLUSION

In this case report we highlighted the importance of distinguishing CRPSt1 and ON especially in stroke patients. It was a chance that the patient does not have central post stroke pain syndrome, because the presence of allodynia and hyperesthesia would cause additional difficulty in differentiation. Early diagnosis and treatment are important in both diseases. A high index of suspicion needed to avoid misdiagnosis of CRPSt1 and diagnostic delay of ON.

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