

Anesthetic Management in a Patient with Mixed Connective Tissue Disease: a Case Report

Miks Konnektif Doku Hastalığı Olan Hastada Anestezi Yönetimi: Olgu Sunumu

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Abstract

The simultaneous occurrence of sytemic lupus erythematosus, systemic sclerosis and polymyositis is denoted as mixed connective tissue disease. The condition involves the gastrointestinal, pulmonary, cardiovascular, renal and locomotor systems and thus presents a challenge for anesthesia. Unilateral spinal anesthesia was administered during right inguinal hernia repair in a 57-year-old man with mixed connective tissue disease. There have been no intraoperative and postoperative complications.

Key words: **Anesthesia; Mixed Connective Tissue Disease.**

Özet

Sistemik lupus eritematozus, sistemik sklerozis ve polimiyositin eş zamanlı ortaya çıkışı, miks konnektif doku hastalığının belirtisidir. Bu durum gastrointestinal, pulmoner, kardiovasküler, renal, lokomotor sistemleri kapsar ve anesteziik açıdanda önem arz eder. Miks konnektif doku hastalığı olan 57 yaşında erkek hastaya sağ inguinal herni onarımı için tek taraflı spinal anestezi uygulandı. İntraoperatif ve postoperative herhangi bir komplikasyon ile karşılaşılmadı.

Anahtar kelimeler: **Anestezi; Miks Konnektif Doku Hastalığı.**

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Introduction

Mixed connective tissue disease (MCTD) was first described by Sharp and co-workers in 1972 (1). It is characterized by high titers of antibodies specific for ribonucleoproteins (anti-RNPAb) and a combination of several defined connective tissue diseases, such as systemic lupus erythematosus, systemic sclerosis and polymyositis.

Approximately 60-91% of the patients are women (2, 3). The most frequent presenting symptoms are arthralgia, Raynaud's phenomenon, puffy hands, myositis, esophageal abnormalities and impaired pulmonary function. Because multiple organ systems are involved, the patients present particular challenges in surgery and anesthesia. Here we report patient diagnosed with MCTD who underwent inguinal hernia surgery emphasizing anesthetic management.

Case report

A 57-year-old man presented to the outpatient clinic of the university hospital with right inguinal pain and swelling of 1-month duration. He was admitted to the general surgery department with the diagnosis of right inguinal hernia. The preoperative examination revealed extensive facial telangiectasias, venous dilation, ronchi in the right lower lung fields and a 3/6 pansystolic murmur on the mitral area. He had had a 5-year history of mixed connective tissue disease and had undergone treatment for anemia. His medical history also included mitral insufficiency, left renal stones, chronic gastritis and operation for cataract. Laboratory investigations revealed mild anemia (hemoglobin: 10.1 g/dL); the urine tested + for protein, ++ for bilirubin and ++ for urobilinogen. The findings of electrocardiography (ECG) were unremarkable. On echocardiography, the left ventricle was found to be dilated; there was 2+ mitral insufficiency; the pulmonary artery pressure was measured as 70 mmHg. The direct radiograph and computed tomography of the thorax showed pleural effusion, infiltrative appearances and cystic images.

Unilateral spinal anesthesia was planned; the patient was informed on the procedure and gave consent. The patient was taken to the operating room. Saline infusion (0.9%) was started. Routine ECG, peripheral oxygen saturation and arterial pressure (noninvasive) were monitored. The patient was placed in the right lateral decubitus position and dural puncture was performed at the L3-L4 interspace with a midline approach. After observing the flow of cerebrospinal fluid through the needle orifice was turned

toward the dependent side and 10 mg of 0.5% hyperbaric bupivacaine was injected without further cerebrospinal fluid aspiration; and the lateral decubitus position was maintained for 10 minutes. After 10 minutes, the patient was turned to the supine position. The degree of motor block on the right side was 3 on the Bromage scale and the level of sensorial block was T9 with the pin prick test.

At operation, the inguinal canal was exposed and a combined direct-indirect (pantaloon) hernia was detected. A primary repair was performed and was buttressed with an onlay mesh graft. The heart rate and blood pressure remained stable throughout the operation (Table I). After the operation, the patient was taken to the recovery room until the level of anesthesia regressed two dermatomes. There was no post operative hypotension, hypertension, bradycardia, tachycardia, nausea and vomiting. The postoperative course was uneventful and the patient was discharged on the postoperative day 3.

Table I. Changes in hemodynamic variable during operation.

	Blood pressure (mmHg)		Heart Rate (beats/min)
	Systolic	Diastolic	
Baseline	110	70	67
After spinal block	110	70	72
1 min	110	70	80
5 min	115	72	76
10 min	112	68	74
15 min	114	67	74
30 min	118	74	78
45 min	116	72	74
60 min	118	70	82
90 min	120	80	76

Discussion

MCTD may involve many organs such as the gastrointestinal, pulmonary, cardiovascular, renal systems. Because multiple organ systems are involved, the patients present particular challenges in surgery and anesthesia. Unilateral spinal anesthesia provided convenient conditions for inguinal hernia repair in a patient with mixed collagen tissue disease.

A variety of connective tissue abnormalities has been demonstrated to be associated with an increased incidence of hernias (4). Abnormal collagen structure and composition as well as fibroblast dysfunction have been

implicated as contributing factors. Connective tissue disorders such as the Ehlers-Danlos syndrome and Marfan syndrome are also associated with an increased incidence of hernias (5,6). However, there is no such evidence of an association between MCTD and hernia formation.

MCTD may involve any part of the gastrointestinal tract. The esophagus is the most frequently involved organ. Heartburn and dysphagia are the most prominent gastrointestinal symptoms. In the majority of the patients (70%), the distal esophagus has no or only low-amplitude peristalsis. Hypotension of the upper esophageal sphincter is also relatively common (7). The pathologies in the esophagus increase the risk of aspiration and aspiration pneumonia under general anesthesia.

The major reason for not preferring general anesthesia in the present case was to avoid the risk of aspiration. Unilateral spinal anesthesia was used in order to decrease the risks of hypotension and bradycardia associated with conventional bilateral spinal anesthesia. Carpenter et al (8) reported hypotension in 33% of the patients and bradycardia in 13% after conventional bilateral spinal anesthesia. Unilateral spinal anesthesia has been shown to cause less prominent cardiovascular effects in comparison with bilateral spinal anesthesia (9).

Asymptomatic pulmonary involvement is present in the majority (85%) of the patients with MCTD. Although pleurisy is frequent, large pleural effusion is rare. Diffuse interstitial infiltrates may be observed on chest x-rays. In some patients, pulmonary involvement may become the predominant problem. In this subgroup, pulmonary hypertension is the leading cause of morbidity and mortality (2,3). Rayes et al (3), in a retrospective study on a 16-year period, detected pulmonary hypertension in 24% of the patients. The patient reported here also had pulmonary hypertension. In the postoperative period, hypoxia, acidosis, hypercapnia may aggravate pulmonary hypertension by sympathetic stimulation (10). Prevention of complications that precipitate hypoxia such as bronchospasm, laryngospasm and respiratory depression should be aggressively avoided. Because we used spinal anesthesia, we did not experience these complications.

Moderately severe anemia and leukopenia may be observed in 30 to 40% of the MCDT cases. Clinically significant Coombs-positive anemia and thrombocytopenia are rare (11). The patient presented here did not have anemia or

thrombocytopenia that could have been contraindicated an operation but had been treated for anemia in the past.

Nephropathy is relatively common and is associated with substantial morbidity and risk of hypertension and chronic renal failure (12). Even in MCTD patients with normal renal function tests, microangiopathy involving the kidneys may be present (13). Therefore, the method of anesthesia and the choice of agents should be conducted to avoid renal toxicity. Spinal anesthesia is an appropriate choice in patients with renal problems because local anesthetics including amide group are metabolized by the liver (14). Moreover, unilateral spinal anesthesia requires a smaller amount of anesthetic in comparison with bilateral spinal anesthesia.

In conclusion, unilateral spinal anesthesia provides convenient operation conditions and hemodynamic stability, does not cause postoperative hypoxia and acidosis that may cause pulmonary hypertension, does not carry the risk of aspiration, has no renal toxicity. Therefore, if the site of the operation is appropriate, it is a good option for MCTD patients.

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