Limited-Form Wegener Granulomatosis Case: Anaesthetic Approach and Literature Review

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Wegener granulomatosis (WG) is a kind of vasculitis that affects small and medium-sized arteries. Necrotizing granulomatous vasculitis of the upper and lower respiratory tracts and necrotizing glomerulonephritis of the kidneys are present. WG affects mainly Caucasian individuals between 15-75 years old, with a mean age of onset of 41 years. It affects both males and females equally. Kidney involvement is not present in the limited form of WG. Peripheral nerve blocks are good alternatives when general anaesthesia is risky. Popliteal block is blockade of the sciatic nerve at the popliteal region. Popliteal block is a kind of peripheral block for surgeries below the knee level. In this article, we report on the anaesthesia management of a 61-year-old limited-form WG patient for whom general anaesthesia was risky because of lung involvement.

Key Words: Wegener granulomatosis, ultrasonography, popliteal block

Introduction

Vasculitis are heterogeneous clinicopathological disorders characterized by inflammation of blood vessels, resulting in destruction and distortion of their walls. Clinical state and categorization hinge on the sizes of the vessels affected. Wegener granulomatosis (WG) is a rare multisystem autoimmune disease that affects small and medium-sized arteries, with unknown etiology (1). Necrotizing granulomas of the upper and lower airways and glomerulonephritis are other components of the disease. Its causes are unknown. It affects mainly Caucasian individuals between 15-75 years old, with a mean age of onset of 41 years (2). It affects both males and females equally.

Lower extremity blocks are frequently used for knee, ankle, and foot operations. Peripheral nerve blocks are very useful when general and/or regional anaesthesia is risky or contraindicated. General anaesthesia side effects and complications may be avoided by applying peripheral blocks in especially risky and emergent cases.

This report presents a case of amputation with WG in a patient for whom general anaesthesia was risky because of pulmonary problems due to WG; central blocks could not be applied because of aspirin use, and anaesthesia was provided by popliteal block.

Case Presentation

A 61-year-old female patient was admitted to the rheumatology department 1 month ago with bloody sputum and redness in her left foot toes. She had been under the care of rheumatology for 5 years with a WG diagnosis. In her arterial magnetic resonance imaging, the proximal left tibial arterial contour was irregular, the middle and distal parts could not be visualized precisely, and other arteries were intact. She was consulted by the orthopaedic and cardiovascular departments. Medical therapy was recommended (clopidogrel, aspirin, cilostazol, methylprednisolone), and she was discharged home. When she was followed up 1 week later, it was seen that the ischemia in the toes had progressed, and she was scheduled for urgent amputation.

Her physical exam showed petechiae on the bilateral upper and lower extremities and inspiratory rales. She had bloody sputum, cough, and hearing loss. Physical examination revealed inspiratory rales and necrosis in the left foot toes 2, 3, and 4. Laboratory findings of the patient revealed hypoalbuminaemia (2.8 gr dL⁻¹), high erythrocyte sedimentation rate (85 mm h⁻¹), anti-nuclear

anticore (ANA) positivity, anti-neutrophil cytoplasmic antibody (ANCA) positivity with cytoplasmic (c-ANCA) pattern, urea 36.8 mg dL^{-1} , creatinine 0.74 mg dL^{-1} , and leukocytosis $20.4 \cdot 10^3 \text{ uL}^{-1}$. Urine test was normal. Posteroanterior chest radiography (X/R) showed bilateral cavities and nodularity (Figure 1). For arterial blood gases, pH was 7.35, pCO_2 was 45 mmHg, and pO_2 was 59 mmHg.

The patient was routinely monitored with electrocardiography, noninvasive blood pressure, and oxygen saturation in the operating room and sedated with 0.03 mg kg^{-1} midazolam. She was turned to the prone position with legs slightly abducted, and her skin was disinfected. The 8-12 MHz linear ultrasound transducer was positioned to identify the sciatic nerve at the subgluteal region (Esaote Mylab 30, Florence, Italy). When the needle (50 mm) tip was confirmed to be adjacent to the nerve, the syringe was gently aspirated, and 0.25% 15 mL bupivacaine was deposited circumferentially. The sensorial blockade onset was at 20 minutes, and the first analgesic requirement was at 8 hours (1 mg kg^{-1} tramadol intravenously). Her written consent was taken for publication of her history before discharge. The patient was discharged on the second day postoperatively without any complications.

Discussion

Wegener granulomatosis is a rare disease characterized by a triad of necrotizing granulomas in the upper and lower respiratory tracts, small and medium-sized vessel vasculitis, and glomerulonephritis. The clinical triad has three components: rhinitis and sinusitis, nodular pulmonary lesions, and renal insufficiency. Vasculitis is both venous and arterial. Many other areas of the body, like the eyes, joints, heart, ears, nervous system, and skin, may also be affected (2). Nasal symptoms include nasal crusting, ulcer, sinusitis, purulent or bloody rhinorrhea, and saddle nose deformity. Eye involvement can lead to visual loss, retrobulbar orbital masses, proptosis, diplopia, conjunctivitis, keratitis, and uveitis. Hearing loss, oral ulcers, subglottic stenosis, cough, wheezing, stridor, and kidney involvement can also be seen. Carrington and Liebow described a form of WG in which renal failure was not found and called it limited-form WG (3). Our patient did not suffer from renal failure either, and so was diagnosed as limited-form WG.

Skin involvement occurs in 35%-50% of patients with WG. Subcutaneous nodules, papules, vesicles, ulcers, petechiae, pyogenic gangrenosum, and Raynaud phenomenon have been reported (4). Our patient had petechiae and purpuric lesions of the upper and lower extremities. She also had upper and lower respiratory tract, ear, and joint involvement.

Patients with WG have 90%-97% multiple or single cavitory lesions in the lungs at diagnosis or during follow-up (5, 6). Chest X-ray shows 34% anomalies; the most frequent ones are bilateral nodular infiltrations, single nodule or infiltrate, cavitory disease, and alveolar haemorrhage (7). Our patient also had cavitory and nodular lesions on chest X-ray (Figure 1).

Figure 1. PA Chest X-ray showing bilateral cavitation and nodularity

The American Rheumatology Society defined four criteria for a diagnosis of WG. These are: 1) abnormal urine sediment, 2) abnormal chest X-ray, 3) oral or nasal inflammation, and 4) granulomatous inflammation on biopsy (1). Our patient fulfills three of these criteria. Although the prevalence in the USA is 0.003%, the prevalence in Turkey is still unknown (5) WG is lethal if left untreated, and the average life expectancy is 5 months in untreated cases. However, response to therapy is very good after diagnosis. A corticosteroid and cyclophosphamide combination results in 93% remission (8). Successful therapy combinations lengthen the life of WG patients; so, we may be faced with WG patients in our routine same-day surgical and anaesthetic practices more frequently. Therefore, we need to know the components of the disease, such as subglottic stenosis, and we must choose the most suitable anaesthetic approach.

Granulomatous inflammation of the upper and lower airways can cause scar formation and subglottic stenosis. This stenosis may cause difficult intubation; so, general anaesthesia should be avoided if it is present. Regional anaesthesia is a good choice in these patients.

Regional anaesthesia consists of central blocks, peripheral blocks, and infiltration blocks. The most important advantage of peripheral blocks is the limitation of the anaesthesia to the nerve's innervation region. Recently, the use of ultrasonography guidance (USG) for peripheral blocks has increased the success rate and decreased complication risks.

Peripheral nerve blocks are used when general anaesthesia is contraindicated or high-risk for both postoperative analgesia and pain therapy (9). Popliteal block is a block of the sciatic nerve at the popliteal region. It is suitable for surgeries below the knee level. Today, with USG guidance, it is easy to apply.

Peripheral blocks are safer than central blocks if the patient is receiving anticoagulant therapy, such as aspirin, mini-heparin, or low-molecular-weight heparin (10). Our patient had been using aspirin for 10 days, so we chose popliteal block as the anaesthesia regimen for her. Peripheral block complications are infection, hematoma, systemic toxicity of local anaesthetic, and neural injury, but these are very rare. We did not see any complications in the patient.

Conclusion

Today, WG diagnosis has become easy with the help of serology, biopsy material pathology, and clinical and radiological evaluation. Peripheral blocks decrease the need for and cost of postoperative intensive care. Using routine same-day anaesthesia practices, with the help of USG, safe and comfortable anaesthesia application is possible for these patients. Peripheral nerve blocks may be good alternatives to general anaesthesia and central blocks for WG patients who require surgery.

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