Spontaneous Spinal Epidural Hematoma in an Adult Patient with Complex Congenital Heart Disease
A Case Report

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Summary
Spontaneous spinal epidural hematoma (SSEH) is considered to be a relatively rare disease that can result in serious neurological sequelae. The pathogenesis and risk factors of SSEH are still unknown, and its differential diagnosis varies widely. Misdiagnosis with more common conditions such as stroke or aortic syndromes can occur. We report the case of a 27-year-old man who developed sudden upper back pain with no specific precipitant. Five days later, he visited our emergency department complaining of weakness in both lower limbs and dysuria. He had a history of intracardiac repair and a Blalock-Park procedure for an interrupted aortic arch and ventriculo-septal defect in infancy. Additionally, he had undergone an aortic root dilatation and aortic valve replacement at the age of 10 because of progression of aortic and supra-aortic stenosis and had received chronic anticoagulation and antiplatelet therapy with warfarin and aspirin, respectively. An emergency spine magnetic resonance imaging scan indicated a mass at the Th3-Th5 level with severe compression of the dural sac and the spinal cord. Emergency excision showed a spinal epidural hematoma. Mild postoperative gait disturbance and dysuria persisted, requiring rehabilitation and intermittent self-urethral catheterization. As patients with adult congenital heart disease have an increased risk of bleeding, they may be at risk of developing SSEH. However, this is the first report to describe such an association.

Key words: Aortic valve replacement, Antiplatelet, Anticoagulant, Aortic valve stenosis, Interrupted aortic arch

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pontaneous spinal epidural hematoma (SSEH) is the accumulation of blood in the spinal epidural space mainly caused by hemorrhage from the spinal epidural venous plexus. Inflammation, edema, and direct compression of the spinal cord result in a variety of neurological symptoms and may lead to severe and irreversible neurological sequelae depending on the size and localization of the hematoma and the time between disease onset and treatment.

SSEH is a very rare condition, with an incidence of 1 case per a 1 million population per year. Although various risk factors, such as use of anticoagulants, blood clotting disorders, arteriovenous fistulas, and hypertension, have been proposed, SSEH can occur without any obvious precipitant, and its pathogenesis remains unclear.

Conversely, the number of patients with adult congenital heart disease (ACHD) has been increasing in recent years. Although ACHDs are associated with a myriad of complications, SSEH has never been reported in this group of patients. In the present article, we report a case of SSEH in an adult man with a history of complex congenital heart disease (CHD). The patient provided informed consent for publication of this report.

Case Report
A 27-year-old man presented to a local clinic with sudden upper back pain during the daytime on July 28, 2015. The attending physicians ruled out cardiovascular disease and prescribed analgesics. On the morning of August 2, he came back to our emergency department with a complaint of weakness in both lower limbs and dysuria.

The patient had been diagnosed with an interrupted aortic arch (type A) and ventricular septal defect at day 6. He had undergone a Block-Park procedure and main pulmonary artery stranguation at day 9 and an intracardiac repair and ascending aortic patch expansion at age 2. Due to progression of aortic and supra-aortic stenosis, a mechanical aortic valve (St. Jude Medical Regent, 21 mm) replacement surgery and aortic root dilatation using the Konno-Nicks procedure were performed at age 10. Subsequently, he received warfarin and aspirin and exhibited a well-controlled international normalized ratio (INR) between 2 and 3. He works as an employee in the general business world and is fully independent. He presented with New York Heart Association class I symptoms. He did not smoke, and he drank alcohol occasionally.

On admission, the blood pressure in the right upper
were palpable and symmetric. Extremities were warm; and peripheral pulses were evident bilaterally. The respiratory and heart sounds were normal; enhanced patellar and Achilles tendon reflexes were present. Body temperature was 36.4°C. The patient complained of numbness extending from the lower limbs up to the abdomen. A manual muscle strength test revealed a decline in strength of the iliopsoas (3/3), quadriceps (4/4), femoral flexors (4/4), tibialis anterior (4/4), and triceps inferior muscles (4/4). Vibration and positional sensations in the lower limbs were decreased. Sensations of touch, pain, and temperature were normal. A positive Babinski sign and enhanced patellar and Achilles tendon reflexes were evident bilaterally. The respiratory and heart sounds were unchanged; extremities were warm; and peripheral pulses were palpable and symmetric.

Blood tests were notable for an elevated INR of 4.38. Chest radiography indicated no congestion, abnormal shading of lung fields, or pleural effusion. Electrocardiography showed a sinus rhythm with no signs of myocardial ischemia. Echocardiography revealed a preserved cardiac contraction and normal aortic valve motion, with no significant valvular, pericardial, or aortic wall disease. Brain and spinal magnetic resonance imaging (MRI) showed a mass shadow in the spinal epidural space between Th3 and Th5, with severe compression of the spinal cord and the dural sac (Figure). The mass had a slightly increased signal with respect to the cerebrospinal fluid on T1-weighted images and an equal signal on T2-weighted images, with a somewhat heterogeneous internal signal. There was no obvious hemorrhage, infarction, or tumor in the head and neck. Based on these findings, a diagnosis of SSEH was made, and the patient was assessed by an orthopedic surgeon. After correction of the INR with vitamin K and fresh frozen plasma, emergency decompression surgery was performed. Heparin was administered immediately after surgery, and warfarin was resumed 2 days after surgery. There was no evidence of aortic valve stenosis on echocardiography. Although the hematoma resembled an arteriovenous fistula macroscopically, histopathology eventually revealed a normal vascular structure. On the second postoperative day, repeat MRI showed no evidence of nerve compression. After starting rehabilitation, the lower limb weakness and paresthesia improved slowly, but difficulty urinating persisted, requiring continued intermittent self-catheterization. No cardiovascular complications occurred during hospitalization, and the patient was discharged from the hospital on postoperative day 94. He has now returned to work and uses a cane only when climbing stairs.

Discussion

This is the first report to describe a patient with CHD who developed SSEH. SSEH presents with a sudden pain in the back and neck that occasionally extends to the extremities. Peripheral manifestations such as flaccid paralysis and poor reflexes are mainly caused by nerve compression, neuronal inflammation, and edema. A time lag between the onset of pain and the appearance of neurological symptoms may be present, ranging from a few hours to several months. Early diagnosis is crucial because irreversible neurological abnormalities may result in death. Additionally, there have been reports of SSEH misdiagnosed as cerebral infarction and treated with antithrombotic therapy, with a consequent risk of serious worsening. Therefore, as it is often difficult to diagnose SSEH based solely on the symptoms, imaging studies are essential.

Two factors might explain the occurrence of this rare complication in our patient. First, prolonged anticoagulation may increase the risk of developing SSEH. Second, CHD may itself be a risk factor for bleeding.

It has been reported that 20%-30% of patients who suffer SSEH are on anticoagulation therapy. Other proposed risk factors include trauma, surgery, arterial malformations, coagulopathy, hypertension, pregnancy, and the Valsalva maneuver. Our patient had been receiving warfarin for approximately 20 years after aortic valve replacement surgery and exhibited a moderately increased INR on admission, which had no clear explanation; this might have temporarily increased the bleeding risk. Several recent studies have found that the range of appropriate anticoagulation for mechanical valves can be reduced. Our patient had undergone multiple cardiac surg-
geries and prostheses since childhood and had been on warfarin and aspirin for a long time. He was at high risk for SSEH and other bleeding disorders. His antithrombotic therapy should be reduced in the future.

The outcomes and life expectancy of complex and severe CHD have improved considerably in recent years; more than 85% of patients with CHD now reach adulthood. A significant proportion of patients with ACHD present with postoperative residual disease and complications that require use of various medications, including anticoagulants. Typical complications include heart failure, cyanosis, arrhythmia, thromboembolism, stroke, liver/kidney disease, and pulmonary hypertension. Additionally, CHD may be independently associated with abnormal hemostasis. In a study by Shebl, et al., abnormal platelet counts, coagulation times, and selectin and platelet factor 4 levels were common in patients with CHD and constituted high-risk factors for both bleeding and thromboembolism. Similarly, Giang, et al. reported an approximately 8-fold increase in the relative risk of hemorrhagic stroke in children and young adults with CHD, compared to matched controls from the general population.

As the number of patients with ACHD increases, it is likely that the occurrence of SSEH will also rise. Despite being a relatively rare condition, SSEH should be included in the differential diagnosis of sudden severe back pain and neurological abnormalities because its management differs considerably from that of cardiovascular disease and stroke, and sequelae may be permanent and severe.

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Disclosure

Conflicts of interest: None.

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