Case Report

Right internal jugular vein thrombosis caused by aneurysm of right-sided aortic arch

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ABSTRACT

A right-sided aortic arch is normally asymptomatic. We report the case of an 84-year-old man with right internal jugular vein thrombosis caused by an aneurysm in a right-sided aortic arch. The patient had undergone open repair of an abdominal aortic aneurysm and had an uneventful postoperative course. However, a routine postoperative contrast-enhanced thoracic and abdominal computed tomography scan showed right internal jugular vein thrombosis. The patient had no history of catheter insertion in the right jugular veins and had no hereditary coagulopathy. It was presumed that the cause of this thrombosis was compression of the right brachiocephalic vein by an aneurysm of the right-sided ascending aorta that was considered too small to require surgical repair. The right internal jugular vein thrombosis was successfully treated with edoxaban.

Learning objective: This case report alerts us to consider that a right-sided aortic arch aneurysm considered too small for surgery can later cause upper-extremity deep vein thrombosis.© 2018 Japanese College of Cardiology. Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Introduction

Right-sided aortic arch is a rare vascular abnormality that is usually asymptomatic. However, symptoms can occur because of the aberrant anatomy, which can lead to compression of mediastinal structures, or because of aneurysm or dissection [1,2]. There have been reports of upper extremity deep vein thrombosis (DVT) caused by a giant aneurysm of the ascending aorta [3,4]. However, there have been no reports of DVT resulting from an aortic aneurysm of moderate size for which surgery is not indicated. We report a case of right internal jugular vein thrombosis in a patient with aneurysm of a right-sided aortic arch.

Case report

An 84-year-old man with a history of Stanford type B aortic dissection was admitted for repair of an infrarenal abdominal aortic aneurysm (AAA). The patient had a history of hypertension and had previously smoked. Chest X-ray on admission showed a right-sided aortic arch (Fig. 1A). Laboratory data revealed a creatinine concentration of 1.4 mg/dL and a D-dimer concentration of 7.2 μg/mL. Preoperative ultrasonography showed no congenital heart anomalies or jugular vein thrombosis.

The patient underwent open surgical repair of the AAA. No catheter insertion in the central veins, including right internal jugular vein, was performed during the perioperative period. On postoperative day 2, the patient began rehabilitation with walking; on postoperative day 4, he began eating solid food. His postoperative course was uneventful. On postoperative day 6, routine chest and abdominal contrast-enhanced computed tomography (CT) scan was performed. Although there had been no thrombosis on the preoperative contrast-enhanced CT scan (Fig. 1B), the postoperative scan showed a contrast defect, indicating a thrombus in the right internal jugular vein (Fig. 1C). It also showed that the left side of the aorta was leaning against the trachea and esophagus, and that a moderate-sized (42-mm-diameter) fusiform aortic aneurysm was compressing the right brachiocephalic vein (Fig. 1D, E). A floating venous thrombus, extending from the mandibular angle to the clavicle, was observed in the internal jugular vein on ultrasonogra-
These findings led to a definitive diagnosis of acute right internal jugular vein thrombosis. The patient had experienced no symptoms such as painful breathing. Because the patient’s body weight was 55.6 kg and his creatinine clearance was 35 mL/min, anticoagulant therapy was started with edoxaban at a dose of 30 mg orally once daily for treatment of jugular vein thrombosis. The patient was discharged on postoperative day 23. D-dimer levels were 7.2 μg/mL at admission, 5.0 μg/mL at the time of CT scan, and 2.5 μg/mL after 1 month of edoxaban treatment. None of the D-dimer levels was normal, however, levels did not increase in the perioperative period.

At 3-month follow-up, ultrasonography revealed that the jugular vein thrombus had totally dissolved without causing pulmonary thromboembolism (Fig. 2B). Oral anticoagulation was discontinued. At 1-year follow-up, the patient had no signs or symptoms of recurrence.

**Discussion**

According to the classification system of Stewart and Edwards [5], malformations of the aortic arch can be one of three types: (i) a mirror-image branching of the normal left arch (the type seen in the present case), (ii) an aberrant left subclavian artery, or (iii) a left subclavian artery that is no longer connected. Another classification scheme of right-sided aortic arch, reported by Knight and Edwards [6], involves two types, according to whether the aorta passes behind the esophagus: right arch with or without retro-esophageal aortic segment, as seen on three-dimensional CT scan. In the present case, the right brachiocephalic vein was being compressed by the aortic aneurysm (Fig. 1D).

The three-stage process of thrombosis has been described by Virchow as hypercoagulability, stasis, and endothelial damage [7]. In the present case, the patient had no history of catheter
insertion in the jugular veins and no hereditary coagulopathy; compression of the right brachiocephalic vein by the aortic aneurysm was presumed to have caused a chronic partial stemming of continuous blood flow. This was likely exacerbated by the patient's prolonged immobility and by hypercoagulability resulting from surgery.

Small to large thrombi of the upper extremities are generally treated with anticoagulation therapy, whereas massive upper extremity DVT is treated with catheter-based intervention or surgical procedures (only if the patient has persistent signs and symptoms or fails anticoagulation therapy)\cite{6}. Because the patient in the present case was asymptomatic, surgical treatment was considered unnecessary, and anticoagulant therapy was given. Therefore, we chose edoxaban, considering the ease of compliance. According to the American College of Chest Physicians guideline\cite{8} concerning patients with acute DVT, initial parenteral anticoagulation is recommended before edoxaban. However, our patient refused intravenous and subcutaneous therapy. Because cases\cite{9} of single-drug therapy using edoxaban have been reported for venous thromboembolism, we chose this treatment. Standard anticoagulation therapy with edoxaban is 3–6 months in duration, so anticoagulation therapy in this case was stopped after 3 months. However, prophylactic use may be necessary when, for example, a patient has undergone a long surgery or is bedbound during hospitalization.

Patients with aneurysm in a right-sided aortic arch require careful follow-up because the right brachiocephalic vein may be compressed by expansion of an aneurysm that is initially considered too small to require surgery. Therefore, when evaluating an aneurysm in right-sided aortic arch, size alone may not determine the need for surgery. Early surgical intervention may be necessary because of the risk of progressive compression of the right brachiocephalic vein with increasing diameter of the aneurysm.

In conclusion, the present case has important clinical implications for treating right jugular vein thrombosis associated with compression of the brachiocephalic vein by an aortic aneurysm in right-sided aortic arch. The case alerts us to consider that a right-sided aortic arch aneurysm considered too small for surgery can lead to upper extremity DVT and may require early surgical intervention.

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