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Staged Management of a Ruptured Internal Mammary Artery Aneurysm

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The rupture of an internal mammary artery (IMA) aneurysm in a patient with type 1 neurofibromatosis (NF-1) is a rare but life-threatening complication requiring emergency management. A 50-year-old man with NF-1 was transferred to the emergency department of Kyungpook National University Hospital, where an IMA aneurysmal rupture and hemothorax were diagnosed and drained. The IMA aneurysmal rupture and hemothorax were successfully repaired by staged management combining endovascular treatment and subsequent video-assisted thoracoscopic surgery (VATS). The patient required cardiopulmonary cerebral resuscitation, the staged management of coil embolization, and a subsequent VATS procedure. This staged approach may be an effective therapeutic strategy in cases of IMA aneurysmal rupture.

Key words: 1. Hemothorax
2. Endovascular procedures
3. Surgery

CASE REPORT

A 50-year-old man with multiple café-au-lait spots on his body was transferred to the emergency department of Kyungpook National University Hospital after the sudden onset of severe dyspnea. We were aware that he had type 1 neurofibromatosis (NF-1) from a previous visit to our hospital 27 years previously, but he had not undergone any treatment. On arrival, his mental status was alert and he had a blood pressure of 119/87 mmHg, but he also exhibited tachycardia, with a heart rate of 130 beats per minute. His initial hemoglobin level was 8.2 g/dL, his hematocrit value was 25.3%, and his platelet count was 67×10^3/L. A chest X-ray showed a massive opacification in the right lung field with a tracheal deviation to the left side (Fig. 1A). A chest computed tomography (CT) scan with medium contrast confirmed a large right hemothorax and extravasation of the contrast medium from an internal mammary artery (IMA) aneurysm (Fig. 1B).

After closed thoracostomy with a chest tube insertion, we removed approximately 1,000 mL of blood. We planned two-stage management for the patient, using endovascular embolization for the proximal portion of the IMA, followed by surgery to evacuate the hemothorax and clip the distal portion of the IMA. Immediately after starting the angiogram, the patient suffered cardiac arrest from hypovolemic shock. During cardiopulmonary cerebral resuscitation with volume replacement, we planned to perform emergency embolization through the right common femoral artery, and emergency angiography

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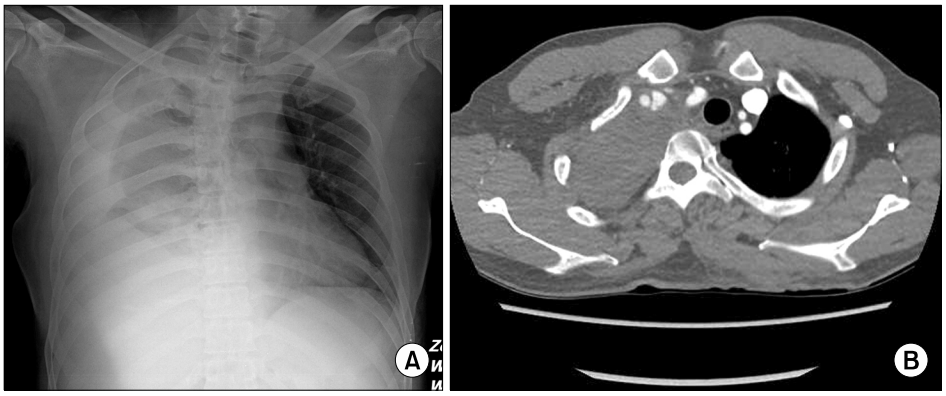

Fig. 1. (A) A chest X-ray confirmed a massive right-sided opacification consistent with a pleural effusion, and tracheal deviation to the left side was observed. (B) Chest computed tomography revealed a right hemothorax and extravasation of the contrast medium from an internal mammary artery aneurysm.

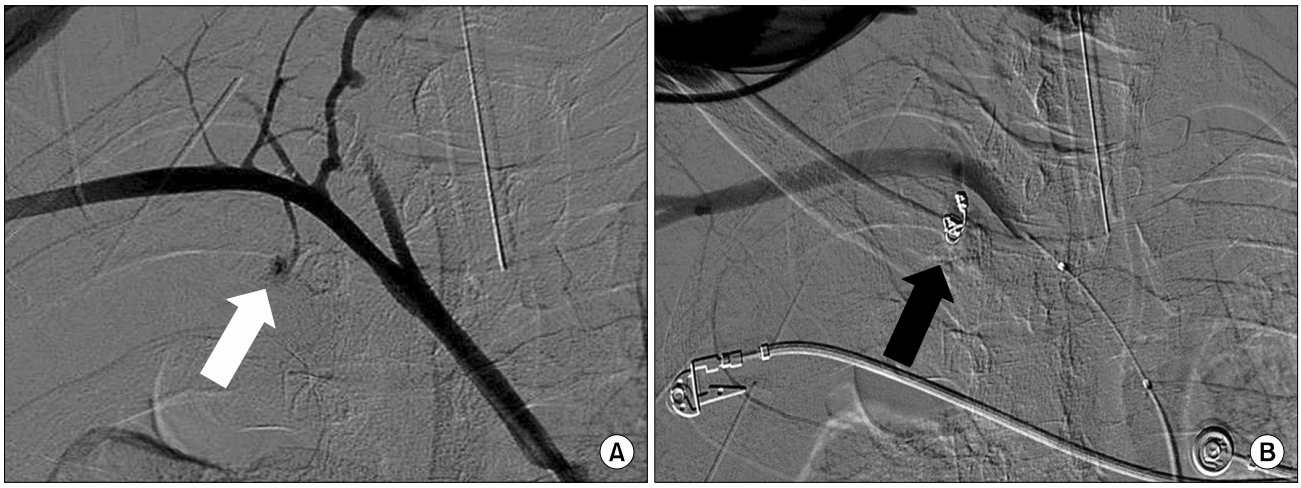

Fig. 2. (A) A catheter was inserted through the femoral artery into the right subclavian artery, and contrast medium was injected. Extravasation was found at the proximal part of the right IMA (white arrow). (B) After coiling was applied (black arrow) to the proximal part of the right IMA, further extravasation was not seen. IMA, internal mammary artery.

revealed a ruptured aneurysm in the right IMA. The interventional radiology team occluded the proximal portion of the IMA with two 5-mm Tornado Embolization Microcoils (Cook Medical Inc., Bloomington, IN, USA) (Fig. 2A, B). Subsequently, superselection of the distal IMA was attempted. However, since the IMA was made discontinuous by an aneurysmal segment, the microcatheter could not reach the distal IMA, making distal embolization impossible. Nonetheless, the patient's hemodynamic status recovered, although his mental state could not be determined at that time. After confirming complete occlusion of the proximal portion of the IMA, the patient was moved to the intensive care unit (ICU) and hypothermic therapy was performed there. In the ICU, the patient was semicomatose and a small amount of bleeding through the chest tube persisted. Serial chest radiographs revealed an increasing pleural effusion that was suspicious for blood in the right lung field.

Three days later, the patient was taken to the operating room to drain the pleural effusion and to clip the distal portion of the IMA. Through a video-assisted thoracic surgery (VATS) procedure, a massive bloody effusion and clot were evacuated. A ruptured mediastinal pleura was observed in the thoracic inlet and a small amount of blood was found to be leaking through the distal portion of the IMA. We clamped the distal portion of the IMA with two hemostatic clips (Fig. 3). After this procedure, the patient's mental state gradually improved and he fully recovered. He was discharged home one month later without sequelae. After six months of fol-

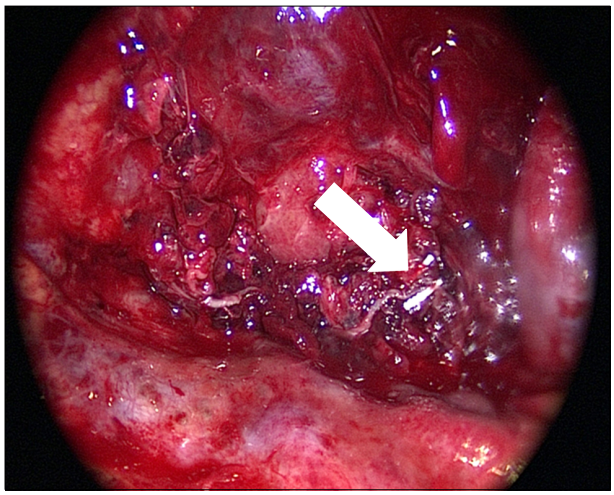

Fig. 3. Operative findings in the right hemithorax. The distal portion of the right internal mammary artery was clipped (white arrow). Further bleeding was not found.

low-up, the patient has remained clinically asymptomatic without recurrent hemothorax.

DISCUSSION

Spontaneous hemothorax caused by a ruptured aneurysm in NF-1 patients is very rare, but can be fatal. Vascular lesions can occur in any vessels, but in the thoracic cavity, ruptures of aneurysms in the intercostal arteries, subclavian artery, and internal mammary artery have been reported in patients with NF-1. The incidence of vascular lesions in patients with NF-1 has been reported to be 3.6% [1]. Hongsakul et al. [2] reported that the most common location of artery rupture was the subclavian and intercostal arteries. Spontaneous hemothorax caused by aneurysmal rupture in NF-1 is very rare, but may be critical, especially if it occurs in a large artery [2,3].

Symptoms vary depending on the size and location of the vascular lesion. Chest pain, dyspnea, and syncope are the presenting symptoms of arterial aneurysmal rupture in the thoracic cavity [4]. In our case, dyspnea occurred as a symptom of a life-threatening case of hemothorax.

Diagnosis requires chest CT or magnetic resonance imaging scans. Furthermore, serial blood tests are needed due to progressive anemia. The diagnosis should be confirmed by selective angiography in order to plan an optimal therapeutic intervention [3,4].

An aneurysmal rupture in a patient with neurofibromatosis should not be corrected by vessel reconstruction. The treatment of choice is surgical or interventional occlusion of the arteries involved. Surgical management is more aggressive and complex, and artery reconstruction is limited by arterial fragility [2-5].

In the case of the rupture of an IMA aneurysm, the treatment depends on the patient's hemodynamic condition. In unstable conditions, emergency exploratory surgery may be needed [4]. VATS is recommended for cases such as ours. Surgical exploration includes removing the hematoma, surgical ligation for bleeding, and packing to achieve hemostasis [6]. However, this surgery is aggressive and problematic due to difficulties in finding the source of bleeding and performing surgical ligation associated with the fragile nature of vascular tissue [7]. Miura et al. [8] reported that they failed to identify the cause of bleeding during surgery, so they terminated the procedure in a patient with an intercostal aneurysmal rupture. In patients who are hemodynamically stable, endovascular treatment is the best choice for arterial rupture in NF-1 patients. Coil embolization or stent grafts are often used in these cases because these treatments are safer and less complicated than surgery [5]. However, fatal rebleeding from the aneurysm or due to backflow from collateral branches has been identified as a possible life-threatening complication after an endovascular procedure [2]. A subsequent VATS procedure may be indicated for cases of aneurysmal rupture in the thoracic cavity to prevent rebleeding and to improve the patient's symptoms. In our case, regardless of whether the patient had been hemodynamically unstable on arrival, we planned a two-stage management strategy that combined endovascular treatment and a subsequent VATS procedure. The interventional radiology team was already prepared when the patient was transferred to our hospital, and we were concerned about possible difficulties in finding the bleeding site through surgery. First, coil embolization of the proximal rupture of the IMA was undertaken, while the distal portion of the IMA was later clipped using two hemostatic clips via VATS.

The prognosis of aneurysmal rupture in NF-1 depends on the patient's status. A large arterial rupture in a patient with

NF-1 is a life-threatening condition that can result in hypovolemic shock and severe dyspnea. Therefore, when encountering an NF-1 patient with a massive hemothorax, early recognition and treatment are necessary [7].

In conclusion, we planned a two-stage management strategy to treat an IMA aneurysmal rupture associated with NF-1. Although the patient required cardiopulmonary cerebral resuscitation, successful results were obtained by coil embolization and a subsequent VATS procedure.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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