Case report - Thoracic non-oncologic

Chest wall bleeding with giant intrathoracic meningocele in neurofibromatosis type 1

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Abstract

We report the case of a 66-year-old woman with neurofibromatosis type 1 who developed chest wall bleeding with severe scoliosis and a giant intrathoracic meningocele. She was brought to the emergency department with acute-onset of left-sided chest pain and clinical signs of hypovolemia. Bleeding control was difficult in the first operation because the tissue was friable and there were multiple subcutaneous bleeding points. During the first operation, the patient developed disseminated intravascular coagulation, which required immediate management; therefore, the surgery was aborted and a repeat surgery was performed later to stop the bleeding. The major cause of bleeding was presumed to be the mechanical stretching of the intercostal arteries and branches of the internal thoracic artery secondary to the severe deformity of the thoracic vertebra and ribs. The massive bleeding remained as a hematoma and did not lead to development of hemothorax. This was believed to be because the giant intrathoracic meningocele supported the expansion of the hematoma and prevented the perforation of the visceral pleura. After the second operation, the hematoma shrunk gradually; however, the patient required ventilatory support because the decrease in the size of the hematoma was accompanied by the expansion of the meningocele.

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1. Introduction

Neurofibromatosis type 1 (NF1), which is also known as von Recklinghausen’s disease, is an autosomal dominant disorder characterized by multiple ‘café-au-lait’ spots, axillary and inguinal freckling, multiple cutaneous neurofibromas, and Lisch nodules in iris. Less common but potentially more serious manifestations of NF1 include plexiform neurofibromas, glioma of the optic nerve and other parts of the central nervous system, malignant peripheral nerve sheath tumors, scoliosis, tibial dysplasia, and vasculopathy [1]. Massive spontaneous hemothorax in patients with NF1 has been occasionally reported as a fatal complication [2]. Intrathoracic meningocele associated with NF1 has rarely been reported [3]. We report the case of a 66-year-old woman with NF1 who developed chest wall bleeding with severe scoliosis and giant intrathoracic meningocele.

2. Case report

A 66-year-old woman was brought to the emergency department with acute-onset of left-sided chest pain and clinical signs of hypovolemia. She reported that six years previously she had undergone shunt placement to the peritoneal cavity for a giant left intrathoracic meningocele and severe thoracic deformity associated with neurofibromatosis (Fig. 1a). On admission, her hemoglobin level was 7.1 g/dl and the coagulation status was stable with a platelet count of 204,000/μl, prothrombin time (PT) of 74.1%, and activated partial thromboplastin time (APTT) of 29.1 s. Enhanced computed tomography (CT) of her chest revealed a huge mass with biphasic density in the left chest wall; the lesion protruded into the pleural cavity and was suspected to be a massive hematoma with active bleeding (Fig. 1b). The large hematoma displaced the intrathoracic meningocele towards the mediastinum. Her thoracic cage was severely deformed (Fig. 2). We performed catheter embolization for the left intercostal arteries (fourth to sixth) and the branch of internal thoracic artery that showed extravasation (Fig. 2). Despite blood transfusion, the anemia had worsened and the hematoma had enlarged the next day (Fig. 1c).

Two days after the patient was admitted, we performed surgery to remove the hematoma and to stop the bleeding. Bleeding control was difficult to achieve because there were many bleeding points and the tissue was friable. The volume of the hematoma and the amount of bleeding loss totaled to 2233 ml. During the surgery, the platelet count dropped from 120,000/μl to 39,000/μl, and the patient developed disseminated intravascular coagulation (DIC). Therefore, the surgery was aborted, and the wound was closed after pieces of gauze packing were placed against the chest wall. Measures were then taken to manage DIC with danaparoid sodium. Four days after her admission, the
The patient underwent surgery again for control of bleeding. During the surgery, coagulation and absorbable hemostatic products did not work well. We finally stopped bleeding by using the technique for spray application of fibrin glue. The total amount of blood loss in this surgery was 4241 ml.

Pathological examination of the specimens obtained from tissue surrounding the bleeding point in the chest wall revealed neurofibroma. After the second operation, the hematoma shrunk gradually; however, the patient required ventilatory support because of persistent chest wall thickness due to repeating surgeries and drainage procedures, and the expansion of the meningocele following the decrease in the size of the hematoma (Fig. 1d). Shunt replacement and adjustment of the shunt flow to shrink the meningocele caused severe intracranial hypotension. Three months after surgery, we attempted to perform cerclage of the meningocele to help improve respiratory dysfunction. However, surgery failed because of severe adhesion between the meningocele and the pleura and bleeding from numerous plexiform subcutaneous neurofibromas despite several measures to maintain hemostasis. The patient finally died of respiratory failure due to the large intrathoracic meningocele and refractory pneumonia eight months after surgery.

3. Discussion

The incidence of vascular lesions in NF1 has been reported to be only 3.6% [4]. Some hypotheses have been proposed previously to explain the rupture of a major artery in NF1. Firstly, the neurofibromatous invasion of the tunica media may reduce the strength of the vessel wall; secondly, neurofibromatous tissue may compress the vasa vasorum of the large artery, resulting in ischemia-induced weakening of a segment of the artery [2]. Thirdly, intimal proliferation and thinning of the media may induce fragmentation of the elastic tissue of the artery [5]. Other theories have also been proposed, such as mechanical stretching of the arteries due to severe scoliosis, tendency of neurofibromas to bleed easy, and dysfunction of hemostasis due to loosening of the skin. In our case, the major cause of bleeding is presumed to be the mechanical stretching of the intercostal arteries and branches of the internal thoracic artery secondary to severe deformity of the thoracic vertebrae and ribs (Fig. 2).

Miura et al. reviewed 12 cases of spontaneous hemothorax associated with NF1 and reported that half of the 10 patients who underwent thoracotomy died due to blood loss during thoracotomy [6]. Shomura and Takahashi reviewed 38 cases of hemothorax associated with NF1 [7].
They reported that, of the 38 patients with hemothorax, 13 were male and 25 were female; the mean patient age was 46 years; and 10 patients had hemothorax on the right side and 28, on the left. The major bleeding site was intercostal artery in 14 cases, subclavian artery in seven; mortality rate in their study was 32%. Although the cause of bleeding in our case and in the abovementioned cases was neurofibromatosis, none of the above cases presented with a chest wall hematoma similar to that seen in the present case. Bleeding control was difficult because the tissue was friable and there were multiple bleeding points. Therefore, we decided to manage DIC first and perform a repeat surgery to stop the bleeding.

Intrathoracic meningocele associated with neurofibromatosis was first reported by Phol [3] and its incidence is very low. Meningoceles are thought to be caused by the herniation of spinal meninges with cerebrospinal fluid extrusion through an enlarged intervertebral foramina. Most patients are asymptomatic [8]. To our knowledge, no report has yet been published on a hydrothorax or hemothorax as a result of the rupture of meningocele, and our patient had the largest meningocele reported to date. On admission of the patient, we considered the possibility of bleeding from the meningocele. However, careful interpretation of the enhanced CT revealed that bleeding originated from the chest wall.

We encountered a case of a rare and severe complication of NF1, namely massive bleeding in the chest wall and intrathoracic meningocele. In this case, the massive bleeding was retained within a hematoma and did not lead to hemothorax. This was believed to be because the giant intrathoracic meningocele supported the expansion of the hematoma and prevented perforation of the visceral pleura.

References