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Title: Surgical intervention for spontaneous hemothorax associated with neurofibromatosis type I

Dr. Ei 18507 Miyamoto corbusier920@yahoo.co.jp MD ¹, Dr. Hiroshi 18508 Hamakawa hamachan@kcho.jp MD ¹, Dr. Ichiro 18509 Sakanoue ichiro.sakanoue@gmail.com MD ¹ and Dr. Yutaka 18511 Takahashi nangpu@kcho.jp MD ¹. ¹ Thoracic Surgery, Kobe City Medical Center General Hospital, Kobe, Japan, 6500047 and ² Cardiovascular Surgery, Kobe City Medical Center General Hospital, Kobe, Japan, 6500047

Body: Rationale: Neurofibromatosis type I (NF1) is a hereditary neurocutaneous syndrome resulted from the mutation in NF1 gene. The clinical diagnosis is comprehensively made by neurofibroma, Café-au-lait spot, osseous deformity and so on. Infrequently, aberrant neurofibromin function results in vasculopathy, which can cause fatal hemorrhage. However, it is unclear how surgical interventions should be made. Case 1: A 53-year-old man with NF1 was admitted for sudden chest pain. Computed tomography revealed left hemothorax and a tumorous aneurysm in the 7th intercostal artery. A thoracic drainage was performed immediately, and a thoracotomy was performed on 18 days after admission. Case 2: A 32-year-old man with NF1 was admitted due to chest back pain and hemodynamic instability. Computed tomography revealed left massive hemothorax and an extravasation from the 9th intercostal aneurysm. An urgent operation via left lateral thoracotomy was performed to remove hematoma and control active bleeding. On 7th postoperative day, the rerupture occurred and was treated with percutaneous transcatheter arterial embolism. Discussion: Management of fatal spontaneous hemothorax associated with NF1 depends on hemodynamic stability. According to 48 reported cases, intercostal and subclavian arteries are popular sites that burst. However, it is often difficult to control, and repeating rupture is known as a fatal complication. In the case 1, surgical intervention was put electively. On the other hand, it was urgently-required in the case 2. Thus, physician should be aware of the possibility of sudden-onset hemothorax in patients with NF1, and surgical intervention might play an important role to treat massive bleeding.