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Video-assisted Thoracoscopic Lobectomy in a Patient with Congenital Factor XI Deficiency

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Abstract

Congenital factor XI deficiency, called hemophilia C, is an autosomal recessive disorder. Bleeding complications are observed in 9.6% of surgeries on such patients. Herein, we presented the first report of a case of lung cancer with FXI deficiency. The patient safely underwent video-assisted thoracoscopic lobectomy after replenishment of coagulation factors *via* preoperative fresh frozen plasma transfusions prior to surgery.

Keywords: Lung cancer • XI deficiency • VATS lobectomy • Hemophilia C • IPMN

Glossary of Abbreviations: FXI: Factor XI; VATS: Video-assisted Thoracoscopic; FFP: Fresh Frozen Plasma; CT: Computed Tomography; APTT: Activated Partial Thromboplastin Time; PT: Prothrombin Time; POD: Postoperative Day; IPMN: Intraductal Papillary Mucinous Neoplasm

Introduction

Congenital factor XI (FXI) deficiency, called hemophilia C, was first reported in 1953 as a mild to moderate bleeding tendency [1]. FXI is not so critical to clotting as other factors, but bleeding is sometimes apparent with FXI deficiency when invasive procedures such as surgery are performed [2]. Such situations call for careful attention and potentially some pre-treatment to avoid life-threating bleeding, depending on the procedure.

The present study is the first report of a patient with FXI deficiency who safely underwent Video-assisted Thoracoscopic (VATS) lobectomy by replenishment of coagulation factors *via* fresh frozen plasma (FFP) transfusion prior to surgery.

Case Report

A 70-year-old Japanese man was referred to our hospital's outpatient clinic due to a chest X-ray abnormality. A chest Computed Tomography (CT) subsequently performed here revealed a solitary 21-mm solid tumor with spiculation in the left upper lobe (Figure 1A). Positron Emission Tomography-CT showed fluorodeoxyglucose accumulation with a Standard Uptake Value of 10.9 at the tumor site without any other place of significant accumulation (Figure 1B). Brain contrast Magnetic Resonance Imaging identified no metastasis. Echocardiography indicated an ejection fraction value of 58% without asynergy. Spirometry showed a 2.55 L/sec forced expiratory volume in one second. Blood chemistry results showed a slightly elevated level of the tumor marker neuron-specific enolase and significant extension of the activated partial thromboplastin time (APTT) at >130 seconds with a prothrombin time (PT) within the normal range. Intrinsic coagulation factor XI was significantly reduced (<1%) while other coagulation factors were normal. From these details, we diagnosed primary lung adenocarcinoma T1cN0M0: cStage 1A3 (UICC Ver 8) with congenital factor XI deficiency.

Four days before surgery, we transfused 4 units (U) of FFP, resulting in an improvement of the APTT to 41.2 seconds. One day before surgery, we transfused an additional 4U of FFP. The surgery was a VATS left upper lobectomy with selective mediastinal lymphadenectomy. The operation time was 165 minutes and bleeding volume was 20mL without intraoperative complications. Macroscopically, a greater than 20-mm whitish tumor was

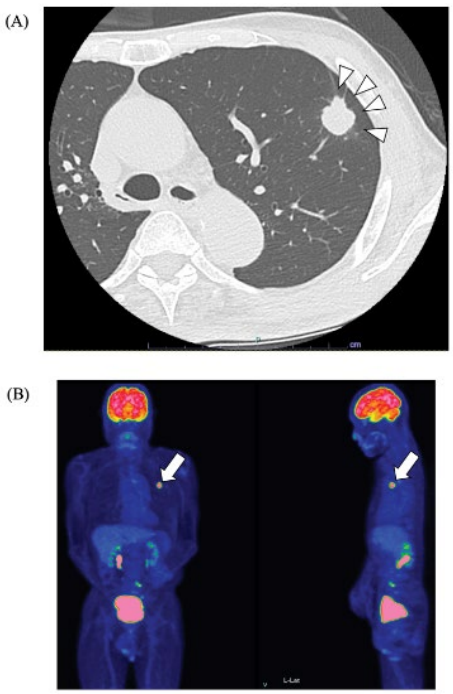

Figure 1. Preoperative image findings. (A) CT image revealed a 21-mm solid lung tumor with speculation in the left upper lobe (arrow heads) and (B) PET-CT indicated FDG accumulation with an SUV of 10.9 at the tumor, without any other place of accumulation (arrowheads).

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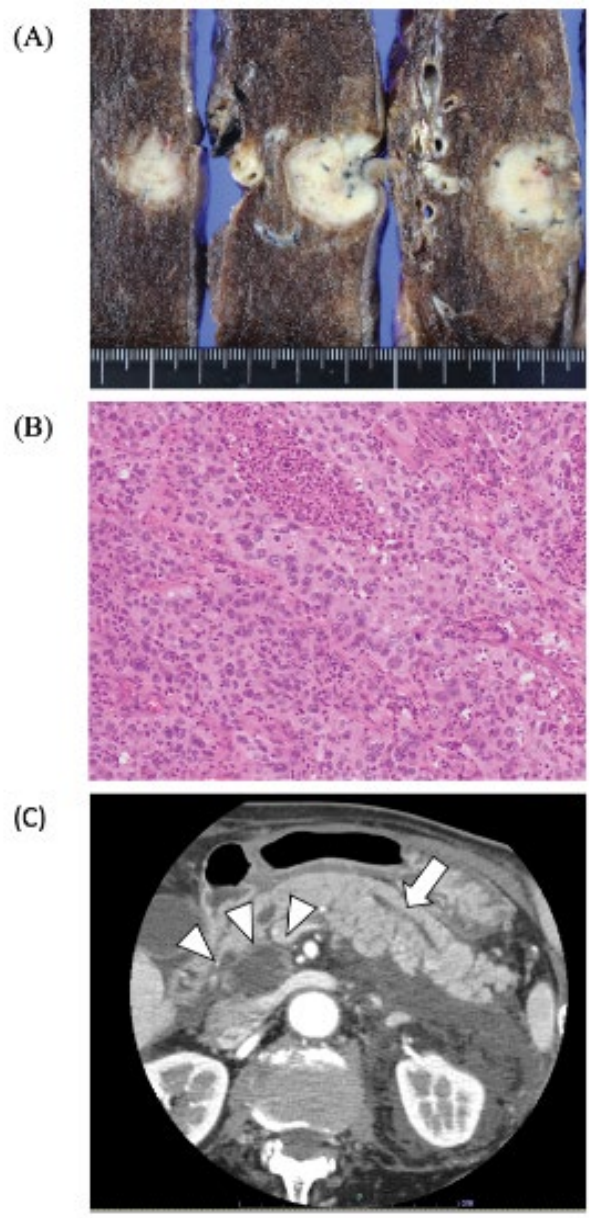

Figure 2. Postoperative findings. (A) Macroscopic split surface revealed a greater than 20-mm solid whitish tumor, (B) Microscopic hematoxylin-eosin findings revealed total size 23 × 18 (invasive size 22 mm) invasive solid adenocarcinoma without lymph nodes metastasis (X200) and (C) Abdominal contrast CT at POD7 revealed a swollen pancreatic parenchyma, a dilated main pancreatic dust (arrowhead), and a cystic mass in the pancreas head (arrowheads), indicating acute pancreatitis due to IPMN.

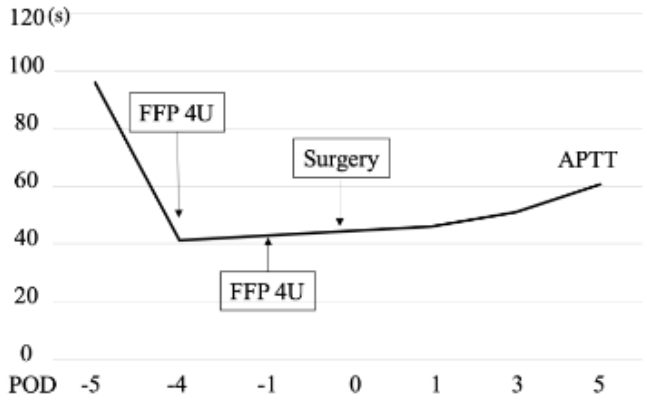

Figure 3. APTT values through the hospital stay. Four units of FFP were transfused at preoperative days 4 and 1.

observed, and microscopic pathological examination suggested invasive solid adenocarcinoma T1cN0M0: pStage IA3 (Figure 2A and 2B).

The postoperative course was uneventful without further transfusion, and the thoracic drain was removed at postoperative day (POD) [2]. However, at POD 7, the patient complained of severe abdominal pain and developed a fever of 38.6 degrees. Serum inflammatory findings and amylase were elevated (white blood cells 23,87 0/μL, C-reactive protein 4.50 mg/dL, amylase 2633 U/L), and abdominal contrast CT revealed a swollen pancreatic parenchyma, dilated main pancreatic duct and ascites around the pancreas reaching to the left kidney. A cystic mass was also observed in the pancreas head, indicating acute pancreatitis due to intraductal papillary mucinous neoplasm (IPMN) (Figure 2C). Conservative treatment with intravenous antibiotic (tazobactam and piperacillin) and nafamostat mesylate improved the symptoms and CT findings. He was finally discharged on POD 24 and has followed up as an outpatient without recurrence.

Discussion

Although thoracoscopic lobectomy in a lung cancer patient with hemophilia is already reported but this is the first report of a VATS lobectomy in a patient with FXI deficiency [3]. Inherited factor XI (FXI) deficiency is a rare autosomal coagulation disorder with an estimated prevalence of 1 in 1,000,000 individuals [2]. Since FXI is part of the intrinsic coagulation system, FXI-deficient patients have a prolonged APTT but a normal-range PT. If an especially prolonged APTT is obtained, FXI activity should be measured along with other coagulation factors. Although the bleeding diathesis in FXI-deficient patients is considerably milder than that in hemophilia A or B, bleeding is nonetheless observed in 9.6% of such patients in the context of surgical procedures including tooth extractions, tonsillectomies, and nasal operations [2,4]. It has been reported that it may be difficult to distinguish severe and mild deficiency on clinical grounds alone, and the lack of a method to reliably distinguish FXI-deficient patients who may bleed from those without such a predisposition has been noted [5,6]. Given the invasiveness of the procedure in our case, it was thought that replenishment of coagulation factors prior to surgery was needed to avoid life-threatening bleeding.

Because human plasma-derived FXI concentrates, which have been shown effective in replenishing XI factor, are not available in Japan, replenishment therapy here is done by FFP transfusion [7]. The main disadvantages of this therapy are the potentials for transmission of infection agents, allergic reactions and volume overload [6]. It is important to confirm preoperative cardiac function to avoid acute heart failure due to volume overload by transfusion.

Because the measurement of serum activated XI factor takes a few days, the APTT value is useful for checking the effect of FFP transfusion. Considering that the half-time of FFP *in vivo* is 3-4 days, we transfused 4U of FFP four days before surgery, and confirmed a recovered APTT value in the normal range, then transfused the same amount one day before surgery (Figure 3). As a result, VATS lobectomy with lymphadenectomy was performed without intraoperative complication.

In our case, acute pancreatitis due to IPMN was observed at POD7. No relationship between FXI deficiency and IPMN or acute pancreatitis has been reported in the literature. Therefore, we assume that this event was independent of the patient's hematologic disorder and recent transfusion status.

Conclusion

Replenishment of coagulation factors by preoperative FFP transfusion can contribute to safely performed VATS lobectomy with lymphadenectomy in patients with FXI deficiency.

Conflict of Interest

The authors have no conflict of interest to declare.

Informed Consent Statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. And the copy of the consent form is available from corresponding author on reasonable request.

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