The patient was a 48-year-old man who had previously been hospitalized due to hemoptysis at 42 years of age.
At that time, a chest radiograph and chest computed tomography (CT) revealed diffuse ground-glass opacity (GGO) in the bilateral lung fields (Fig.1,2), and a bronchoscopic examination revealed the accumulation of large amounts of blood in the trachea and bronchi (Fig.3).
The patient's bronchoalveolar lavage fluid (BALF) was bloody and contained numerous hemosiderin-laden macrophages.
A culture test of the BALF revealed no findings.
An electrocardiogram and transthoracic echocardiogram revealed no abnormalities.
A questionnaire that was completed at that time did not reveal the patient's hemorrhagic episode or a family history of bleeding disease, with the exception of his younger brother who had been diagnosed with hepatitis C.
Furthermore, he had not received any drugs that had the potential to cause DAH.
Following the above-mentioned examinations, DAH due to some sort of vasculitis was suspected.
Thus, treatment with high-dose intravenous methylprednisolone (1,000 mg daily) for 3 days followed by prednisolone (25 mg daily) was initiated.
This relieved his symptoms.
Chest CT showed the resolution of the GGO.
The patient's prednisolone dose was tapered and eventually discontinued at two-and-a-half years after his discharge from our hospital.
The patient was readmitted to our hospital with a recurrence of hemoptysis at 46 years of age.
The above-described therapy was initiated and led to the improvement of his condition.
The prednisolone dose was tapered from 60 mg daily to a maintenance dose of 5 mg daily.
At 48 years of age, he was readmitted to our hospital with a further recurrence of hemoptysis.
At this point, he had steroid-induced diabetes mellitus, which was treated with glimepiride (3 mg daily).
At admission, his weight was 75 kg and height 167 cm; his vital signs were as follows: blood pressure, 178/102 mmHg; pulse rate, 109 beats/min, pulse oximetry, 97% in room air; and body temperature, 36.8°C.
Chest auscultation revealed fine crackles in the right lung field.
No skin rash, subcutaneous bleeding or joint swelling were present.
A chest radiograph and CT showed the presence of diffuse GGO in the bilateral lung fields.
Mild anemia was observed (hemoglobin, 11.3 g/dL), although hemoglobin levels had been 14.6 g/dL prior to the hemoptysis episode.
The activated partial thromboplastin time (APTT) was prolonged to 53.5 seconds (normal range, 25.1-40.7 seconds).
Laboratory tests showed that the patient's blood glucose and hemoglobin A1c levels were 299 mg/dL and 8.2%, respectively, due to the steroid-induced diabetes mellitus.
Autoantibodies for various collagen diseases were negative (Table).
We diagnosed the condition as a recurrence of DAH and again administered high-dose intravenous methylprednisolone for 3 days, followed by prednisolone (60 mg daily).
His condition improved, as had been observed during the previous episodes.
A further detailed inquiry regarding the patient's medical history revealed that, as an elementary school student, he had been hospitalized and had received blood transfusions twice following abnormally heavy bleeding after tooth extraction; however, a specific congenital bleeding disorder had not been diagnosed at that time.
It was also revealed that his younger brother had been diagnosed with hemophilia B during adolescence.
Considering the possibility of hemophilia, his blood coagulation factors were examined, revealing that his factor IX activity was 3%.
The patient was subsequently diagnosed with moderate hemophilia B.
By the time of this diagnosis, the patient's DAH had already resolved with the corticosteroid therapy.
We decided to continue treating the patient using prednisolone alone, without coagulation factor IX replacement therapy.
No recurrence of DAH or hemorrhagic symptoms have been observed during 3 years since the tapering and discontinuation of prednisolone.