

Two cases of patients with thoracic SMARCA4-deficient undifferentiated tumors with severe airway stenosis who underwent airway stenting

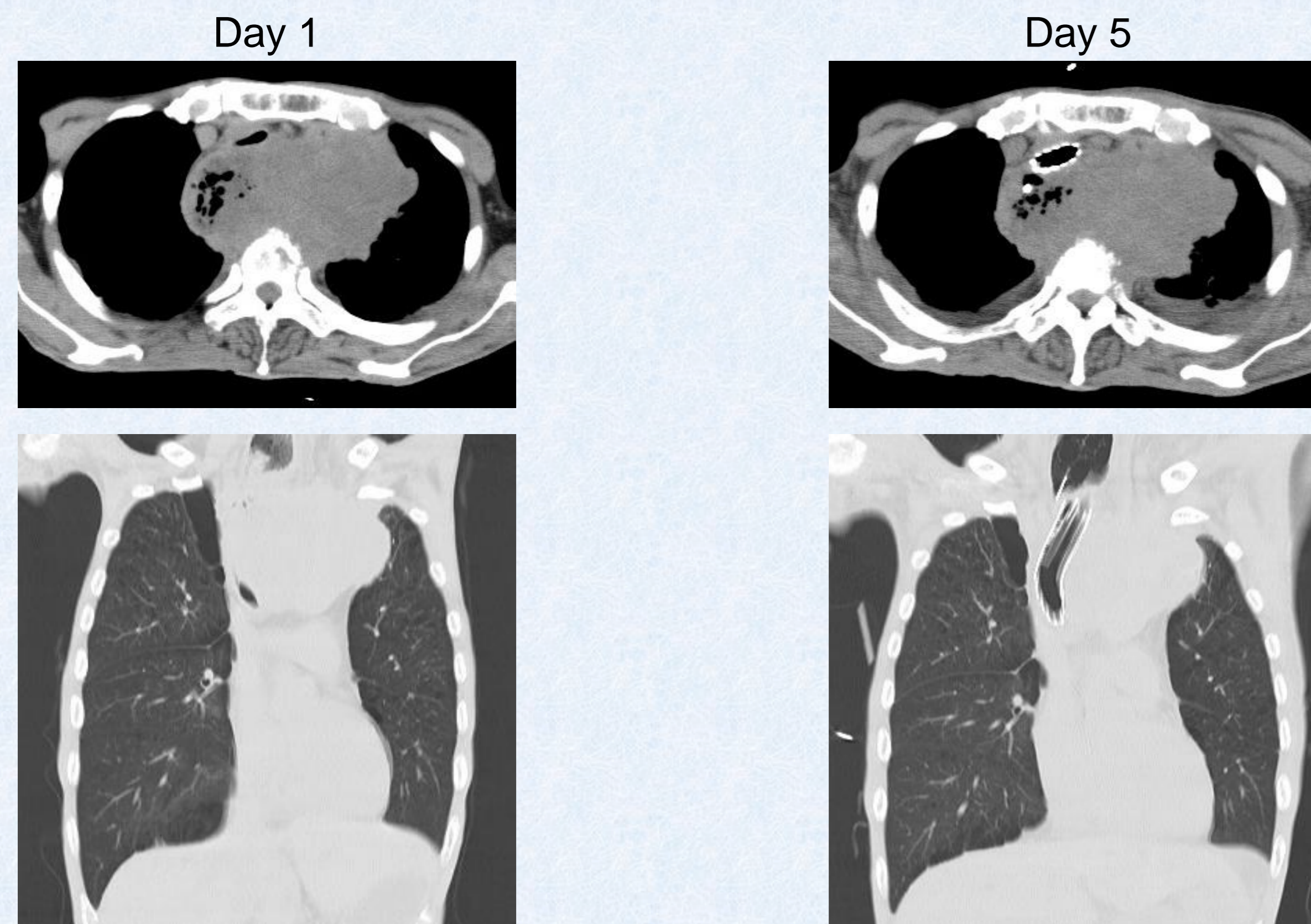
Y. Shinohara, Y. Ishii, A. Iwakoshi, M. Oki
National Hospital Organization Nagoya Medical Center, Nagoya, JAPAN

Background

Thoracic SMARCA4-deficient undifferentiated tumors (SMARCA4-UT) are a rare and relatively new disease that was first reported in 2015. These tumors frequently occur in the mediastinum and pulmonary hilum, and there is no established treatment. The tumors progresses quickly, and patients often have a poor prognosis.

Case 1: A 58-year-old man

He had dyspnea for two months prior to admission and underwent emergency transport to the previous hospital for exacerbated dyspnea. His past medical history was remarkable for a prior intracranial hemorrhage, symptomatic epilepsy, shoulder arthritis and a 64.5 pack-year history of smoking. He was transferred to our hospital for bronchoscopic treatment of a mediastinal tumor that was found to cause an airway stricture by CT scan. On admission to our hospital, his ECOG performance status score was 4 and his SpO₂ was 96% on O₂ 10L/min via reservoir mask.



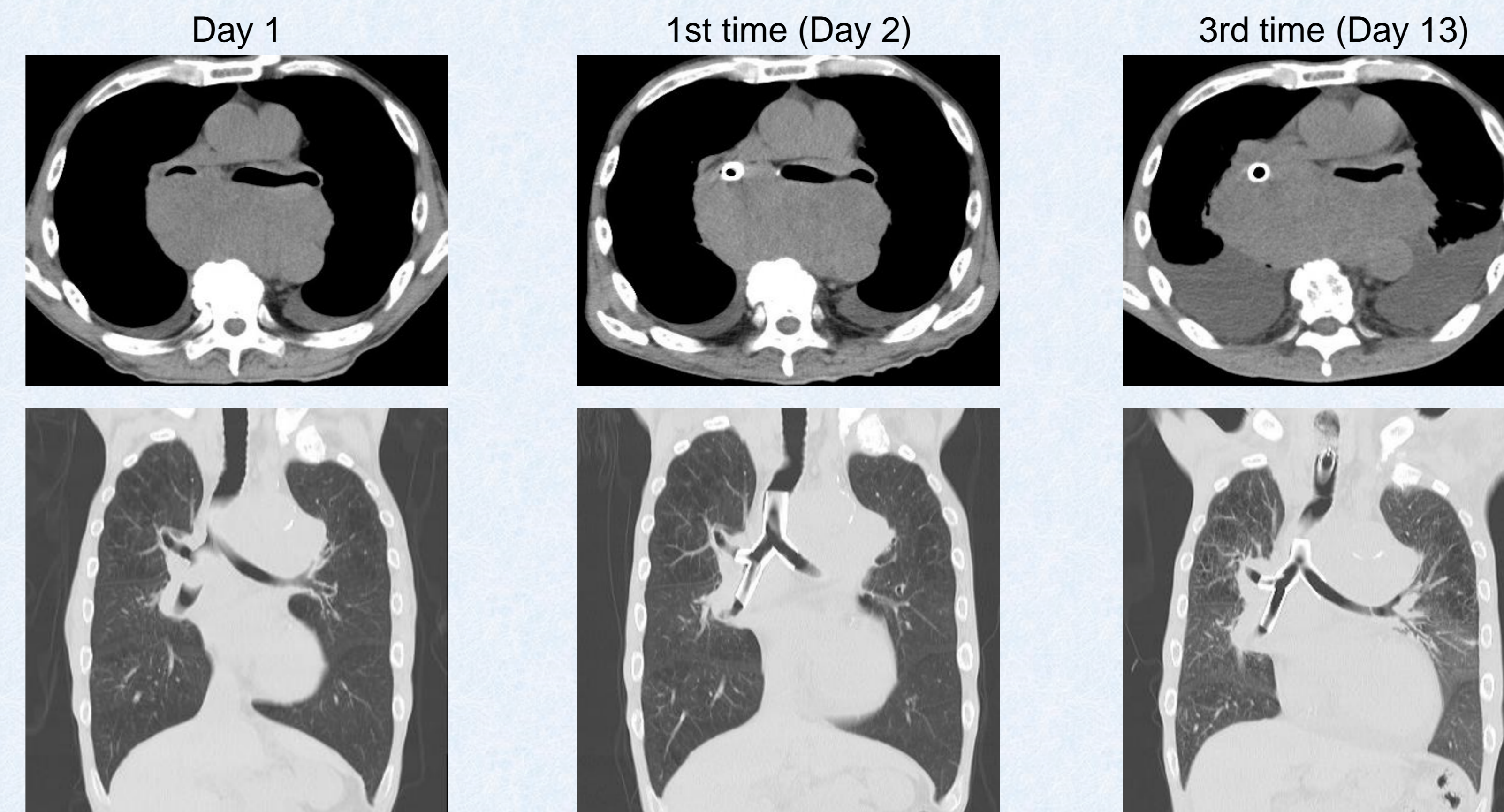
Day 1
He underwent airway stenting using a rigid bronchoscope under general anesthesia. We placed an Ultraflex® (18 x 80 mm) stent for the tracheal stricture and secured it with 2 stitches. A Mini-Trach® was also inserted because expectorant failure was predicted. After airway stenting, his SpO₂ was 97% on room air and his dyspnea was resolved.

Day 14
He was diagnosed with a SMARCA4-UT by biopsy taken from the esophagus on day4 and he was delegated to receive best supportive care due to his performance score being 3.

Day 22
He died due to rapid disease progression.

Case 2: A 64-year-old man

He was admitted to his previous hospital for close examination of a mediastinal tumor. He was transferred to our hospital for bronchoscopic treatment, because he had extensive tracheal stenosis from the trachea to both main bronchi, and increasing dyspnea on exertion. His past medical history was significant for a previous cerebral aneurysm, ureteral calculi, appendicitis, and a 44 pack-year history of smoking. On admission to our hospital, his ECOG performance status score was 3 and his SpO₂ was 96% on room air.

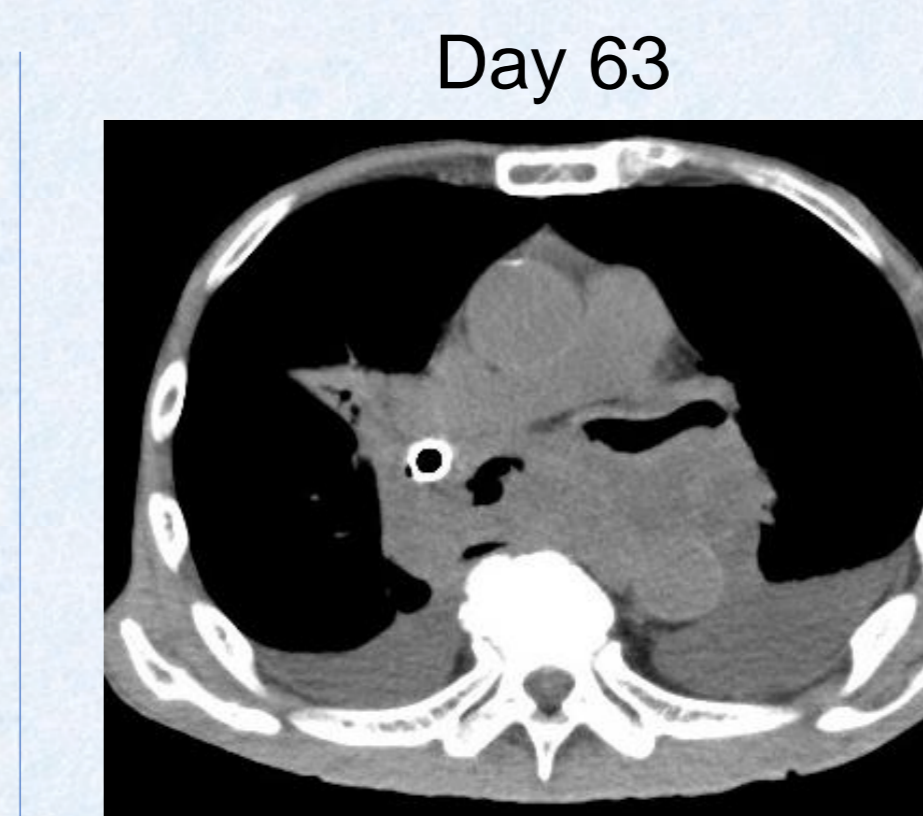


Day 2
He underwent airway stenting using a rigid bronchoscope under general anesthesia. We placed two Dumon™ Y-shaped silicone stents.
▶14-10-10: Right main bronchus 10mm, right upper lobe bronchus 10mm, bronchus intermedius 25mm.
▶16-13-13: Tracheal 40mm, left main bronchus 15mm, right main bronchus 10mm.
After airway stenting, his dyspnea on exertion disappeared.

However, he repeatedly failed to expectorate and desaturated. His performance status score worsened to 4. He underwent stenting on days8 and on day 13.

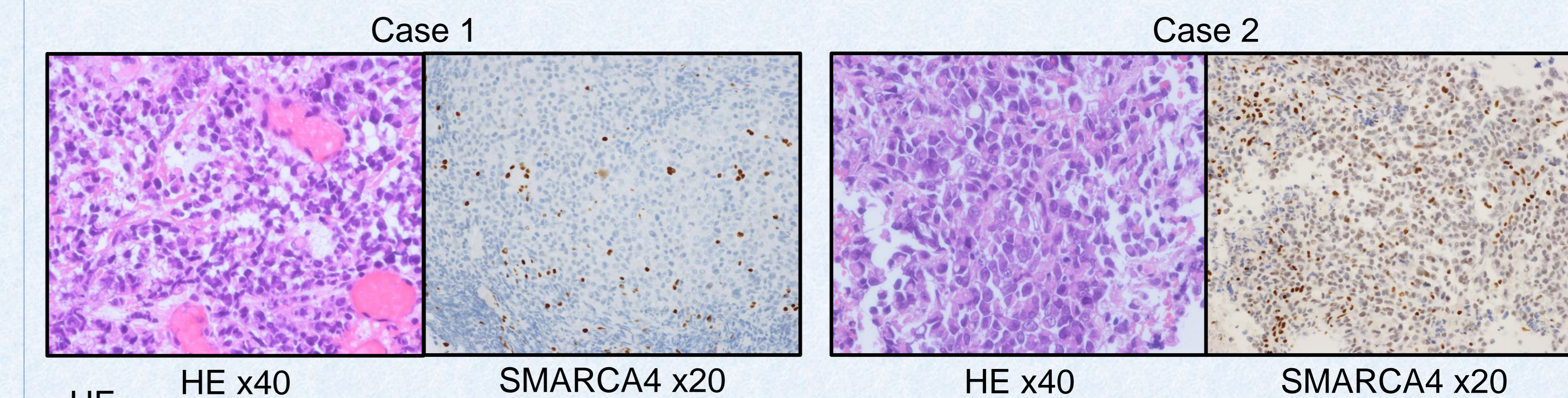
Day 13
He underwent a 3rd airway stenting procedure using a rigid broncho scope under general anesthesia. We placed two Dumon™ Y-shaped silicone stents.
▶14-10-10: Right main bronchus 10mm, right upper lobe bronchus 10mm, bronchus intermedius 25mm.
▶16-13-13: Tracheal 25mm, left main bronchus 15mm, right main bronchus 10mm.
And performed a tracheotomy because expectorant failure was predicted.

Day 18
He was diagnosed with a SMARCA4-UT by intraoperative biopsy, and was deposed to radical radiation therapy.



Day 63
The radiation therapy resulted in tumor shrinkage and his performance status score improved to 3. He was delegated to receive the best supportive care because his performance status score was 3, and he was transferred to a convalescent hospital at day 83.

Day 126
He died due to disease progression.



▶HE
These cases show similar pathologic features, consisting of relatively monotonous, variably discohesive round to epithelioid cells with component of rhabdoid cells.
▶SMARCA4 (BRG1)
The tumor cells show diffuse loss in case 1 and diffuse severe reduction in case 2 with normal cells serving as internal controls.
▶Another immunohistochemistry
Pan-keratin(very focal+), claudin4(very focal+), TTF-1(-), p40(-)

Discussion

- ▶SMARCA4-UT typically affects younger patients from age30 to 59.¹
- ▶ Cases are predominately in males. (male:female = 9:1)^{2,3}
- ▶The most common site of involvement is the mediastinum followed by the pleura and the lungs.^{1,3}
- ▶Overall median survival averages 4 to 7 months (range 1– 13 months), and 2 years overall survival rate of 12.5%.¹
- ▶A few case reports have shown that SMARCA4-UT may benefit from treatment with immune checkpoint inhibitors or platinum-based chemotherapy.^{4,5}

Conclusion

We present two cases of patients with SMARCA4-UT with severe airway stenosis that required airway stenting. SMARCA4-UT frequently occur in the mediastinum and pulmonary hilum, and severe airway stenosis can be fatal. However, stenting may enable subsequent treatment.

References

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