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LETTER TO THE EDITOR

Is the case really a SMARCA4-deficient thoracic sarcoma?

To the Editor,

I read with great interest the recent publication of the report by Takada *et al.*[1] With regard to this case challenge, I have some concerns about the diagnosis which was made.

First, SMARCA4-deficient thoracic sarcoma (DTS) has been reported to have unique clinical features. It is more common in men, more common in heavy smokers (≧20 pack-years), is a relatively large tumor (mean size; 9.2 [2.2–18.3] cm) with mediastinal involvement, and often has emphysema in the lung fields.[2] What about the smoking history of the present case? Was there any emphysema in the lung field? Based on the chest images presented (Fig 1 in Reference 1), the tumor was relatively small and mediastinal involvement was not apparent. Subcutaneous metastases are different from bone, adrenal glands, and intra-abdominal lymph nodes, which are typical metastatic sites of SMARCA4-DTS.[2] The clinical picture is somewhat atypical of SMARCA4-DTS. Could you advise about the smoking history of the patient and other clinical information?

Second, in the presented case, the expression of SMARCA2 was retained. However, the expression of SMARCA2 is lost in about 80% of SMARCA4-DTS cases, and the expression of Caludin-4 is reported to be lost even when SMARCA2 is maintained.[2] Additional immunostaining information such as Claudin-4, SALL4, CD34, TTF-1, p40, and cytokeratin may be needed to confirm the diagnosis.[2-4] I am afraid that it is not possible to diagnose SMARCA4-DTS only with immunostaining, as presented in the report.

SMARCA4-DTS is characterized by clinical and immunopathological profiles and must be distinguished from SMARCA4-deficient non-small cell lung carcinoma (NSCLC). Because of its rapid progression and poor prognosis, many readers are interested in effective treatments for the disease. I believe that this case report is extremely meaningful and strongly request that the accuracy of the diagnosis of SMARCA4-DTS be ensured.

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References

1 Takada K, Sugita S, Murase K *et al.* Exceptionally rapid response to pembrolizumab in a SMARCA4-deficient thoracic sarcoma overexpressing PD-L1: A case report. *Thorac Cancer* 2019; **10** (12): 2312–5.
2 Rekhtman N, Montecalvo J, Chang JC *et al.* SMARCA4-deficient thoracic sarcomatoid tumors represent primarily smoking-related undifferentiated carcinomas rather than primary thoracic sarcomas. *J Thorac Oncol* 2020; **15** (2): 231–47.
3 Yoshida A, Kobayashi E, Kubo T *et al.* Clinicopathological and molecular characterization of SMARCA4-deficient thoracic sarcomas with comparison to potentially related entities. *Mod Pathol* 2017; **30** (6): 797–809.
4 Kunimasa K, Nakamura H, Sakai K, *et al.* Patients with SMARCA4-deficient thoracic sarcoma and severe skeletal-related events. *Lung Cancer*. 2019; **132**: 59–64.