Rare lung cancers—2021 update on challenges, advances, and opportunities

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In the diagnostic and treatment landscape of various types of lung cancer, rare tumors represent a major challenge for patient management in terms of their biology, classification, and clinical behavior. In this scenario, the term rare lung cancer refers to a multitude of heterogeneous diseases including primary pulmonary sarcoma (PPS), neuroendocrine neoplasms (NEN) of the lung, thymic epithelial tumors (TET), malignant pleural mesothelioma (MPM), and other tumors often characterized by diagnostic pitfalls, the unavailability of prognostic and predictive biomarkers, and the lack of standardized treatments. In this issue, we compiled articles on the above-mentioned rare disease and tried to elaborate clinical dilemmas and to shed some light on diagnostics, treatment, and clinical management of rare lung cancers.

Dr. Metovic and team give a pathological update on NEN of the lung and highlight the importance of the diagnostic workup and incorporates news from the forthcoming 5th edition of the World Health Organization (WHO) classification of lung NENs. Furthermore, the authors give some hints to biomarkers, which should be given consideration in clinical practice [1].

Next Prof. Junraithmayr presents the double-edged sword of immune checkpoint inhibition (ICI) treatment in advanced TET [2]. Patients with TET are known to experience higher rates of severe immune-related adverse events, as the thymus is a major regulator of self-reactivity, however also an attractive target for ICI therapy.

MPM is a very difficult to treat malignancy and shows in advanced stage a dismal prognosis due to the aggressiveness of the disease. Dr. Absenger et al. discusses the new treatment advances with incorporation of dual ICI blockade and gives an outlook on combination therapies improving overall survival in MPM [3].

Another rare primary lung tumor is PPS. Dr. Nagl presents the complex and individual interdisciplinary management of a patient with PPS and tries to increase the awareness for PPS [4].

Driver mutations in non-small cell lung cancer as anaplastic lymphoma kinase (ALK) translocations have distinct treatment algorithms and have revolutionized our daily practice. Now rare resistance profiles to targeted therapies are emerging and challenging us daily. Dr. Hempel and colleagues present a case with occurrence of a G1202R ALK resistance mutation after treatment with alectinib and response to lorlatinib [5].

The present issue summarizes recent advances in rare lung cancers, including in part practice-changing data regarding novel diagnostic and therapeutic approaches based on biological insights that continue to contribute to significantly extend oncologists' clinical management portfolio and improve outcomes. These efforts and consequent positive developments in an orphan disease field can be attributed to the highly active multidisciplinary thoracic oncology community and I am already looking forward to the next advances and highlights.

Conflict of interest A. Pircher declares that he has no competing interests.

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