Editorial

Metastatic only primitive neuroectodermal tumor: smoke without fire can be more dangerous

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Multiple, diffuse, well circumscribed large pulmonary nodules as an incidental finding is pathognomonic for disseminated cancer. For all cancers, the lung is the second most common site of metastasis after the liver (1,2) and the most common for Ewing Sarcoma family of tumors (ESFT) (3). Disseminated disease at presentation signifies poor prognosis regardless of the cancer origin, however both the treatment regimen and treatment response as well as the ultimate outcome is highly dependent on the type of metastatic cancer, and thus the first step is to obtain tissue diagnosis. While less invasive strategies always appear to be less morbid, they can be non-diagnostic and prolong time to treatment. Definitive surgical biopsies are invasive but conclusive and, despite slightly higher morbidity, end up being more effective.

The pathologist's role in investigative oncology cannot be underestimated in these difficult cases. In the case presented, the pathologists were able to determine the presence of the *EWSR1* leading towards further investigation of an osseous primary. The possible location of interosseous primary based on PET-CT and MRI, while certainly unlikely, is potentially more plausible in the adult patient population with what seems like a wider variety of disease characteristics with higher rates of soft tissue primaries and unconventional locations based on a review of patients over 50 (4). Treatment analysis in adults has many limitations relating to the lower numbers in an already rare disease, however the rates of disease free and overall survival are similar to those found in children (5-7). Regardless of primary location, the mainstay of treatment in metastatic disease, as was sought by the authors, is systemic with potential addition of whole lung irradiation. The relatively small size of the primary tumor with limited primary metastatic disease to the lung alone carries a better prognostic indicator than additionally disseminated disease (8,9).

Interestingly, there is importance to be placed on long term follow up of the pulmonary lesions and further assessment by thoracic surgery after completion of chemotherapy. In both the pediatric and adult populations there is wide variability in the response rate of pulmonary metastasectomy dependent on the type of cancer (10). For the Ewing sarcoma pediatric population, the data is minimal and mixed, leading to no clear-cut role for metastasectomy at this time (11-13). Careful consideration of both patient and tumor factors is required for optimal patient selection when considering pulmonary metastasectomy (14).

The authors should be congratulated on their investigative work towards finding a finite diagnosis for their patient and should be charged with keeping good follow up to provide all viable options for treatment based upon treatment response. Long term follow up would be quite interesting as it may end up revealing the primary lesion.

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