Editorial

Primitive neuroectodermal tumor with lung metastases and an unknown primary: a unicorn?

Adam J. Bograd

Division of Thoracic Surgery, Swedish Cancer Institute, Seattle, WA, USA

Correspondence to: Adam J. Bograd. Division of Thoracic Surgery, Swedish Cancer Institute, Seattle, WA, USA. Email: Adam.Bograd@swedish.org.

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"*When one hears hoofbeats, think of horses, not zebras*," is a common line taught to medical students and residents throughout the world. Rarely however those hoofbeats belong not to horses or zebras, rather to unicorns.

In the journal *Current Challenges in Thoracic Surgery*, Mizelle and colleagues present a case report which describes the first documented case of metastatic Ewing sarcoma (or peripheral primitive neuroectodermal tumor) to the lung with an unknown primary (1). The case described is very atypical for presentation of a Ewing sarcoma. While overall rare, Ewing sarcoma is most commonly identified in the pediatric patient population; adult cases are exceedingly rare (2). Furthermore, in instances of metastatic disease there is typically an easily identifiable primary lesion. Most commonly this is an osseous primary within the pelvis or the lower extremities, but sometimes within the chest wall (3). Ewing sarcoma typically possesses an aggressive phenotype with at least ¼ of patients presenting with metastatic disease. It follows that patients presenting with locally aggressive disease generally outlive their counterparts who present with synchronous metastases. Multimodality treatment, including system chemotherapy, has been associated with improved overall survival (4).

Despite overlap between the treatment algorithms of some soft tissue sarcoma subtypes (i.e., type of chemotherapy used), there are subtle differences and nuances associated with the treatment of rare sarcoma subtypes. The authors demonstrate a key principle in sarcoma management: a thorough histopathologic evaluation to confirm the sarcoma subtype and initiate proper systemic therapy. Accurate diagnosis of Ewing sarcoma is crucial as treatment for this disease carries with it both short- and long-term toxicities (3). Unfortunately, despite early initiation of systemic therapy, cure rates are fairly poor among patients that present with metastatic Ewing sarcoma (4).

Mizelle and colleagues are to be commended for this contribution to the thoracic surgical literature as it highlights several key points in the diagnosis and management of Ewing sarcoma:

(I) correct diagnosis is paramount as proper systemic treatment may depend on the sarcoma subtype;
(II) outside consultation with a sarcoma specialist should be pursued if any uncertainty of diagnosis;
(III) multidrug chemotherapy is the foundation of therapy for metastatic Ewing sarcoma.

While this is the first report of metastatic Ewing sarcoma to the lungs without identification of a primary lesion, thoracic surgeons should be aware of the potential for lung metastases from an identifiable Ewing sarcoma primary. With more standardization of the pathologic evaluation of Ewing sarcoma, namely by immunohistochemistry and fluorescence in situ hybridisation (FISH) to identify *EWSR1* rearrangements, there may be increasing identification of this sarcoma subtype in the adult population. As such, thoracic surgeons should be aware of Ewing sarcoma, its diagnosis, prognosis, and treatment, as they may encounter this disease in some fashion during their careers.

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