Ewing Sarcoma with Intradural Metastatic Disease:

An Unusual Adult Course of a Common Pediatric Cancer

Matthew Erlich M.Phil[1], Christopher Terry MD[1,2], Emmanuel Apor MD[1,2], Eric Winer MD[1,2]

The Warren Alpert Medical School of Brown University, Providence, RI[1], Rhode Island Hospital, Providence, RI[2]

INTRODUCTION

Ewing Sarcoma is a highly malignant bone and soft tissue tumor. The most common age of presentation is the second decade of life, with only 10% of patients presenting at greater than 20 years of age.[1] The tumor can develop anywhere in the body, with the most common sites including long bones, the pelvic girdle, and ribs. Metastatic disease to the lungs, bones and bone marrow is commonly seen. By contrast metastatic disease involving the CNS is rare.[2]

The following describes a 25 year-old male presenting with vertebral body Ewing Sarcoma metastatic to the dura.

CLINICAL VIGNETTE

In January 2014 a 25 year-old right-handed Caucasian male with a history of myotonic dystrophy presenting with four days of upper thoracic pain, ascending parasthesia and bilateral lower extremity weakness. An MRI at that time revealed a T3-T4 vertebral body lesion involving the right pedicle with extraosseous extension into the T3-T4 neural foramen. Neurosurgery was consulted for surgical decompression. Posterior laminectomy provided stabilization, and pathology revealed Ewing Sarcoma (Fig. 1, 2). Further staging was negative. Chemotherapy treatment and subsequent consolidative radiation therapy reduced the tumor burden, and MRI six months later revealed stable post-radiation changes at T1-T4 with no evidence for recurrent or progressive tumor.

In August 2014 the patient represented with five days of altered mental status and multiple episodes of urinary incontinence. The initial laboratory work-up was unremarkable, but an MRI of the brain revealed five new enhancing, dural based lesions, most consistent with metastatic Ewing Sarcoma concerning for intra-axial invasion (Fig. 3).

A Head CT and spot EEG showed stable bifrontal vasogenic edema and he was noted to be aphasic. On August 6, Neurosurgery performed an emergent left craniotomy for debulking and resected a large blue, suckable tumor described as not clearly extra-axial, with intradural bone growth (Fig. 4). He required 9 days of tube feeding. His course improved and he remained hemodynamically stable, demonstrating slow but continuous neurologic recovery based on exam, most notably in regard to motor function and aphasia. He completed two weeks of postoperative adjuvant whole-brain radiation.

The patient represented one month later with upper respiratory symptoms and pancytopenia. Bone marrow biopsy showed dysplasia and infiltration of Ewing's. Patient's course was complicated by numerous aspiration events and two weeks after admission, the patient was deceased due to cardiac arrest secondary to hypoxic respiratory failure in the setting of pneumonia.

FIGURES

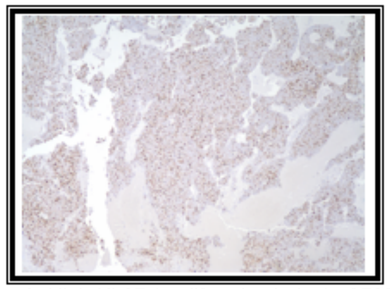

Figure 1: High Ki-67 (proliferative index) of 70% in tumor cells

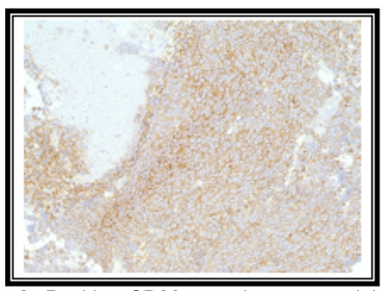

Figure 2: Positive CD99, membranous staining of tumor cells

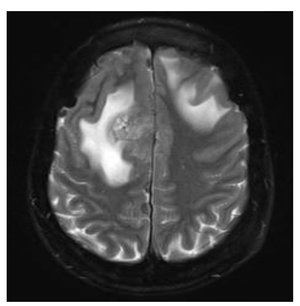

Figure 3: T2 weighted, dural based enhancing lesions with associated underlying vasogenic edema

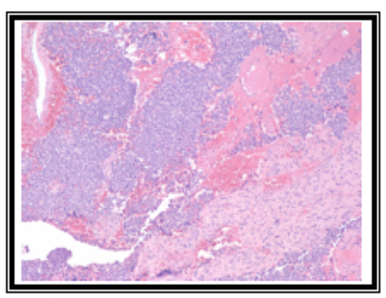

Figure 4: Small round blue cell tumor (cells with high nuclear cytoplasmic ratio) with hemorrhage and adjacent desmoplastic change of the dura

DISCUSSION

This case is notable for the relatively advanced age of presentation and the extremely rare presence of intradural metastatic disease. We found only three published cases in English medical literature featuring intradural metastatic disease.[3,4,5] A common feature of among each was an initial presentation after the age of 20.

Treatment guidelines are available for primary tumors and more common sites of metastasis, but there is no literature directed on therapy for CNS metastases given its rare occurrence.[6] The rarity of CNS involvement would argue against preventative radiation, but these cases raise important questions about screening and long-term surveillance in adult survivors of pediatric Ewing's, as well as those who present in adulthood. Prolonged life expectancy for individuals with Ewing's will likely increase the prevalence of CNS metastases, thus further exploration of treatment guidelines for CNS involvement may be warranted.

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