Primary intradural extraosseous Ewing’s sarcoma of the lumbar spine presenting with acute bleeding

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Abstract

We report the case of a 28-year-old female with primary extraosseous Ewing’s sarcoma who presented initially with a low back pain and a right S₁ radicular pain. Before scheduled surgical removal, she suddenly developed an unusual complication of an acute hemorrhage and an acute cauda equina syndrome. Emergency surgery was done which demonstrated an acute bleeding. Treatment was followed by the chemotherapy and the adjuvant radiotherapy. Follow-up has been done for 6 years after presentation.

Keywords: Ewing’s sarcoma; lumbar spine; hemorrhage

Introduction

Extraosseous Ewing’s sarcoma (EES) is an extremely rare, primary soft tissue tumor that has been grouped along with the peripheral primitive neuroectodermal tumor and Ewing’s sarcoma of the bone under the Ewing’s sarcoma family of tumors.1,2 It commonly affects adolescents or young adults and most commonly involves the chest wall, lower extremities, pelvic and retroperitoneum.1–3 In the spine, EES usually occurs in the epidural spaces or the paravertebral area.2,3 Primary intradural EES in the spine is extremely rare and only seven cases have been reported in the literature.1–3 We report a case of primary EES associated with a spinal nerve root which has been presented with an acute bleeding.

Case report

A 28-year-old female presented with a three-month history of low back pain and right S₁ radicular pain. Magnetic resonance imaging (MRI) of the lumbar spine revealed a well-defined intradural mass lesion at L5–S₁ level, isointense on T₁-weighted, and hypointense on T₂-weighted images with enhancement after contrast media administration. The lesion was closely related to the right S₁ nerve root with intradural foraminal extension (Fig. 1a–c). CT scan of the lumbar spine and sacrum was normal. Elective surgery was planned for her. Prior to admission for the elective surgery, she suddenly developed severe low back pain which was followed by progressive distal lower limbs weakness and urinary retention. An emergency laminectomy of L5 and S₁ was performed and after dural opening, a highly vascular dark red tumor with prominent blood vessels and hemorrhagic area was found. There were some blood clots around the tumor with subarachnoid hemorrhage. The tumor and blood clots were removed totally. Histopathological examination revealed a small round blue cell tumor (Fig. 1d). Immunohistochemical study was positive for S₁₀₀, synaptophysin, CD₉₉ and glycogen (PAS; periodic acid-Schiff) and was negative for EMA, GFAP, and CD₂₀. Fluorescent in situ hybridization analysis of the tumor sample showed chromosomal translocation t(11; 22) (q₂₄; q₁₂).

Based on histopathological findings and immunohistochemical analysis and lack of bone involvement, the final diagnosis was EES. Cranial, cervical and thoracic spine MRI, whole body bone scan, and bone marrow biopsy were normal. The treatment was continued by five cycles of chemotherapy with vincristine (2 mg/m²), doxorubicin (75 mg/m²), and cyclophosphamide (1200 mg/m²) alternating with ifosfamide (1800 mg/m²) and etoposide (100 mg/m²). Then, the adjuvant radiotherapy was delivered in the amount of 50 Gy to the lumber region. Chemotherapy with the above-mentioned drugs continued for another 12 cycles after the radiotherapy. She had complete neurological recovery. After 6 years of follow-up, no tumor recurrence has been found.

Discussion

EES is a highly malignant, small round blue cell tumor, which is derived from the neural crest cells.1 Spinal nerve root may be the origin of intradural EES as in our case. Common clinical presentations of intradural EESs are radicular pain and progressive sensory and motor disturbance.1–3 There has
only been one other report of acute presentation due to the hemorrhage.3 The EES diagnosis is based on the basis of histological and immunohistochemical findings with no evidence of bony involvement during the time of presentation.1–3 EES is a systemic disease, even if documented metastases are not found during the time of diagnosis. Thus, a combined-modality treatment is mandatory. The mainstay of the treatment is the complete surgical excision of the tumor, if possible, followed by an adjuvant chemotherapy and radiotherapy.1–3 Our patient was treated with the surgery, radiotherapy and a total of 17 cycles of chemotherapy. She had no recurrence after 6 years.

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References