Primary extraosseous Ewing sarcoma of the cervical epidural space

Case illustration

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Extraosseous Ewing sarcomas (EESs) are rare malignant neoplasms of the soft tissue that do not arise from bone and have histological characteristics similar to osseous ESs and peripheral primitive neuroectodermal tumors.1,2 Extraosseous ESs are usually found in the lower extremities and paravertebral regions in patients in their 20s and 30s.3,4 Most primary EESs extend into the anterior epidural space through foraminal widening and bone destruction;4 involvement of only the cervical epidural space is highly unusual, although EESs with both intradural5 and extradural3 settling have been reported.

This 28-year-old man presented with progressively increasing neck pain and gait disturbance. Imaging of the cervical spine revealed a well-circumscribed, posterior epidural mass compressing the spinal cord and extending from C-3 to C-5 without foraminal widening (Fig. 1A and B). The patient underwent a C3–4 laminoplasty and gross-total resection of the lesion (Fig. 1C). Immunohistochemical studies revealed a malignant round-cell tumor consistent with osseous ES (Fig. 2), which had no primary focus. The patient’s symptoms resolved immediately after surgery. The patient underwent a chemotherapy regimen that included ifosfamide (2500 mg/m2 for 3 days), mesna (2500 mg/m2 for 3 days), and doxorubicin (60 mg/m2 for 1 day). This was repeated six times at 21-day intervals. Radiotherapy targeted the spine between C-2 and C-5 with a fractionated total dose of 4500 cGy (180 cGy/day for 25 days). No evidence of recurrence or metastasis was found during 18 months of follow up.

Due to its similarity to other lesions, establishing the diagnosis of EES is difficult1 but should nonetheless be considered in the differential diagnosis of spinal epidural tumors. Because all lesion types cannot be identified on the basis of imaging studies alone, a histo pathological examination must be conducted to establish a definitive diagnosis.1,2 Early, aggressive resection combined with chemo- and radiotherapy can produce good results in patients with EES.1,2

References