Primary spinal peripheral primitive neuroectodermal tumor

Epidemiology

Primary spinal peripheral primitive neuroectodermal tumors (pPNETs) are extremely rare entities that predominantly occur in children and young adults.

Treatment

Microsurgical GTR of the tumor is the preferred method of treatment. Radiotherapy plays an important role in improving the prognosis of patients with pPNETs. GTR combined with radiotherapy and chemotherapy may be the best treatment modality [1].

Outcome

Spinal PNETs, like their cranial counterparts, are aggressive tumors and patients with these tumors typically have short survival times despite maximal surgery, chemotherapy, and radiation. Because no standard management guidelines exist for treating these tumors, a multitude of therapeutic strategies have been employed with varying success [2].

Case series

The clinical data of 24 patients, who had been surgically treated from April 2003 to February 2018 in Department of Neurosurgery, Tongji Hospital, Wuhan, in whom immunohistochemical staining results had confirmed the diagnosis of primary spinal pPNETs, were retrospectively analyzed. To analyze the factors related to prognosis, the Kaplan-Meier method was used for univariate analysis, the log-rank method was used to test the significance of difference, and multivariate analysis was performed using Cox regression.

The overall 1-year, 2-year, and 5-year survival rates were 73.2%, 48.1%, and 12.0%, respectively. The median survival time (MST) of all patients was 21 months. Univariate analysis showed that the extent of tumor resection, adjuvant radiotherapy, and chemotherapy were the factors influencing patient prognosis after surgery (all $P < 0.05$); sex, age, tumor location, and preoperative Karnofsky performance scale (KPS) scores were not the influential factors for prognosis of patients after surgery (all $P > 0.05$). Multivariate analysis showed that gross total resection (GTR) of tumors and adjuvant radiotherapy were independent factors influencing the prognosis of patients with pPNETs (all $P < 0.05$).

Primary spinal pPNETs are extremely rare, and they have a poor prognosis. Microsurgical GTR of the tumor is the preferred method of treatment. Radiotherapy plays an important role in improving the prognosis of patients with pPNETs. GTR combined with radiotherapy and chemotherapy may be the

best treatment modality [3].

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13 patients (nine females and four males) with primary intraspinal pPNETs who were surgically treated from April 2008 to February 2014. Histopathologic findings revealed the expression of CD99 in all cases. Limb weakness was the most common initial symptom (11/13, 85 %). The tumors were located mainly at the cervical level (6/13, 46 %) and in the epidural space (10/13, 77 %). The radiological diagnosis was neurinoma or meningioma in most cases (10/13, 77 %). Gross total resection was achieved in 77 % (10/13) of patients. During a mean follow-up of 25.5 months, local relapse occurred in 8 (61.5 %) patients and distant metastases occurred in 8 (61.5 %) patients. The overall 1-year survival rate was 77 % (10/13), and the overall 2-year survival rate was 54 % (7/13). The 2-year survival rate was 57.1 % in patients with adjuvant chemotherapy and 50 % in those without chemotherapy. Gross total resection and adjuvant radiotherapy with or without chemotherapy demonstrated a longer survival period (1-year survival rate: 100 %; 2-year survival rate: 86 %). The data showed that primary spinal pPNETs are extremely rare and aggressive tumors with a poor prognosis. Radical resection is advocated. Gross total resection combined with adjuvant radiation may help to significantly improve patient survival period. Chemotherapy may also help to slightly prolong patient life [4].

Three patients of 8, 9 and 18 years of age, who presented with variable grades of neurological deficit were diagnosed as having a dorsal intramedullary lesion, a holocord lesion and cervical extradural tumor with extraspinal extension, respectively, and were operated at our institute. The histopathology of all 3 children revealed PNET. The clinical course, image characteristics and outcome of the 3 children are described, and the relevant literature is reviewed. The following conclusions were drawn from the present study and review of the literature. PNET may manifest itself as a primary lesion of the spine unlike the more common drop metastases from an intracranial lesion. PSPNET may be intramedullary, intradural and extradural with variable extraspinal extension. PSPNET may present as holocord intramedullary lesion, an entity which has not been described earlier. These lesions have a short history, significant neurological deficits and rapid course of illness. PSPNET, though an established entity, did not find a place in the WHO 2000 classification of CNS tumors. Hence its status has to be define [5].

Case reports

A 26-year-old male presented with progressive low back and lower limb pain for 1 month. Based on MRI and histopathological findings, he was diagnosed with primary intramedullary PNET. The patient was treated two times with microsurgical resections. Follow-up visit at 14 months after the first surgery showed that the patient is neurologically intact and free of disease. PNETs should be considered in the differential diagnosis of an intramedullary spinal cord tumor manifesting as progressive neurological deterioration [6]

A 5-year-old Moroccan boy, who presented with torticollis for 1 month. Computed tomography scan and Magnetic resonance imaging of the cervical spine revealed an extradural, dumbbell-shaped mass

with extra-spinal extension at the left C1-C6 level. Multiple biopsy specimens were obtained. Histological examination revealed a highly cellular neoplasm composed of diffuse sheets of tumor cells having monomorphic, round to oval, finely vesicular nuclei. Immunohistochemical findings confirmed the diagnosis of intraspinal peripheral primitive neuroectodermal tumor [7].

A two years old female child presented with weakness both lower limbs. Preoperative MRI of the spine and paravertebral region Iso - hyper intense posterior placed extradural lesion, non contrast enhancing from D11-L2 levels with cord compression D9 to L3 laminectomy done. Granulation tissue found from D11 to L2. with cord compression. The granulation tissue removed in toto. The pathological findings were consistent with PNET. Post operative neurological improvement was minimal. Cranial screening ruled out any intracranial tumour. Hence a diagnosis of primary spinal PNET was made [8].

A 18-year-old female with conus intramedullary tumor diagnosed to be primary spinal primitive neuroectodermal tumor following histopathological examination after surgery. The diagnosis of such a tumor is very crucial as the management strategies for these are relatively unclear and are associated with a poorer outcome compared to the other common intramedullary spinal tumors [9].

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