

## CASE REPORT

# PRIMARY INTRASPINAL PRIMITIVE NEUROECTODERMAL TUMORS: REPORT OF A CASE

Hamed Reihani-Kermani MD\*, Batool Amizadeh MD

*Department of Neurosurgery, Kerman University of Medical Sciences, Kerman, Iran*

**Background** – Primitive neuroectodermal tumors (PNETs) are rare neoplasms that are usually seen in children and frequently metastasize in the central nervous system. We present a case of primary intraspinal PNET.

**Methods** – A 22-year-old woman presented with back pain and leg weakness. Magnetic resonance imaging of the spine revealed an intradural extramedullary lesion at the level of thoracic vertebra 12. Total tumor removal was performed and pathology was consistent with a PNET.

**Results** – Within 4 weeks, there was a recurrence of symptoms with paraparesis at the original site and at the level of C1, T6, and T9. The PNET responded dramatically to radiotherapy.

**Conclusion** – To date, only 14 cases of primary intraspinal PNET have been reported. The presented case is the second reported, in which the tumor was intradural and extramedullary. Intraspinal PNETs have a poor prognosis with reported cases surviving less than 2 years.

**Keywords** • primitive neuroectodermal tumor • radiotherapy • spinal neoplasm

### Introduction

Primitive neuroectodermal tumors (PNETs) are malignant small cell neoplasms, mainly occurring in children but can occur at any age.<sup>1</sup> They most commonly occur in the cerebellum, but can also arise in the pineal gland, cerebrum, spinal cord, brain stem, and peripheral nerves.<sup>2</sup>

To date, only 14 cases of primary intraspinal PNETs have been reported.<sup>2-11</sup> Of these cases, only one has been intradural and extramedullary;<sup>7</sup> the remainder have been intramedullary,<sup>2,4</sup> extradural,<sup>10</sup> or arose from the cauda equina.<sup>6,8,9</sup> In this case report, a primary intradural extramedullary PNET is described.

### Case Report

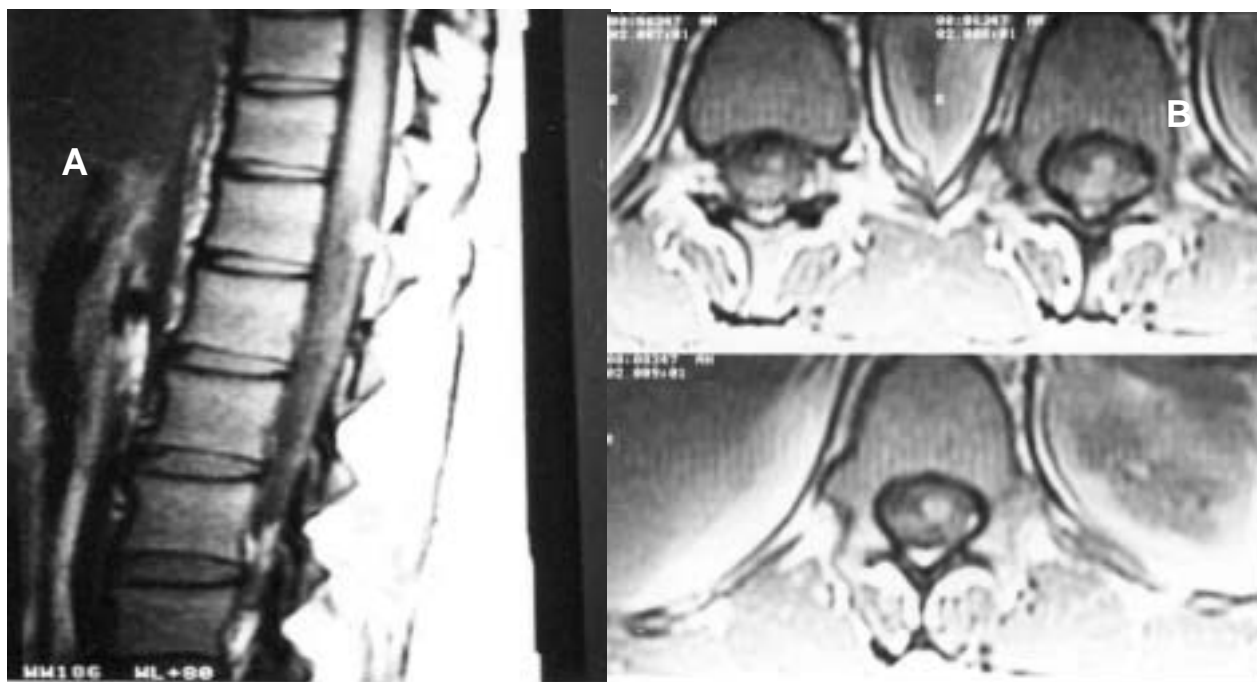
A previously healthy 22-year-old woman presented with back pain, radiating to the right leg, of 1 month duration. She had experienced progressive weakness of her right foot 5 days before admission.

Any disorder of the urinary sphincter was not noted. A neurologic examination revealed weakness on extension of the right knee, right foot dorsiflexion, hyperesthesia on the lateral and medial aspects of the leg, and dorsum of the foot. Joint position sense was absent at the large toe on the right side. Deep tendon reflexes were hypoactive. General physical examination was normal. Spinal magnetic resonance imaging (MRI) revealed an intradural extramedullary space occupying lesion, in the posterior part of the spinal canal at the level of T12 (Figure 1).

A laminectomy was then performed at level of T12. On opening the dura, an extramedullary tumor was found. The tumor was well demarcated. Total tumor removal was performed under the operating microscope. In the early postoperative period, the patient had complete resolution of all neurologic deficits and pain.

Histologic examination of the mass revealed a highly cellular tumor, consisting of mainly small round to oval cells with hyperchromatic nuclei and remarkably scanty cytoplasm. Homer-Wright pseudorosettes were also present (Figure 2). Immunohistochemistry was performed using synaptophysin, neuron-specific enolase, glial

\*Correspondence: H. Reihani-Kermani MD, Department of Neurosurgery, Bahonar Hospital, Kerman, Iran. Fax: +98-341-2150215, E-mail: [H-Reihani@hotmail.com](mailto:H-Reihani@hotmail.com).



**Figure 1.** A and B. Preoperative sagittal and axial spine MRI show an intradural extramedullary space occupying lesion at the level of T12.

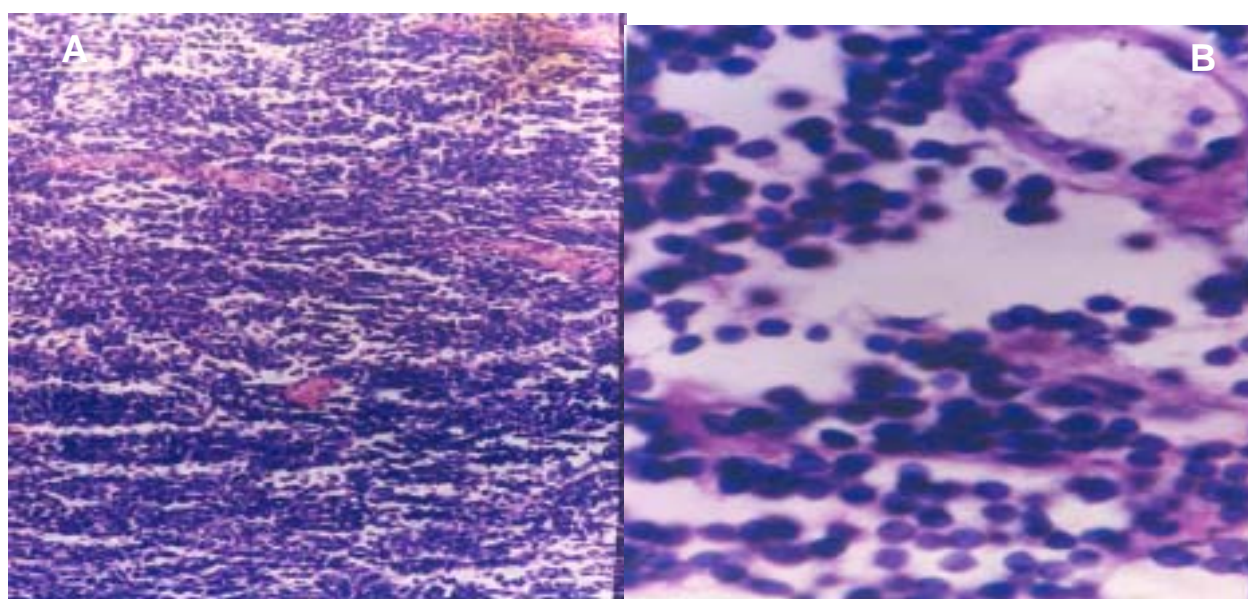
fibrillary acidic protein (GFAP) and cytokeratin. Tumor cells showed immunostaining with synaptophysin and neuron-specific enolase (Figure 3) but were negative for GFAP and cytokeratin.

The diagnosis of PNET was made. The patient was recommended for further investigations but refused.

Four weeks later, the patient began to experience recurrence of back pain and leg weakness. Neurologic examination revealed

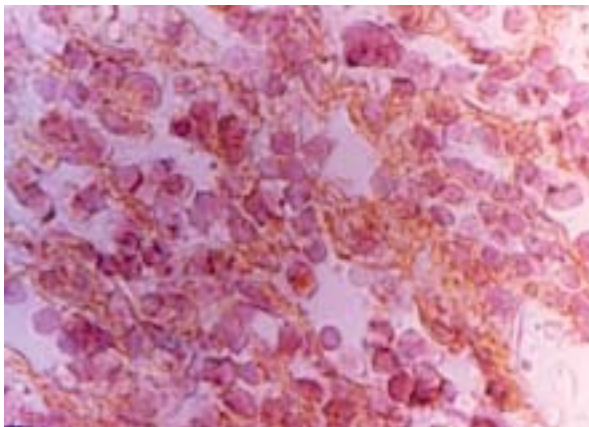
weakness of both legs, sensitive to pinprick at T8 and bilateral hyperactive deep tendon reflexes. Postcontrast MRI of the spine and head showed small tumor recurrence at the original site and three other masses at the level of C1, T6, and T8-T9 (Figure 4). MRI of the skull did not show any other tumors (Figure 5).

The patient was scheduled for craniospinal radiation and chemotherapy. During radiation, the patient's neurologic condition improved. Eight



**Figure 2.** A (left) and B (right). A poorly differentiated tumor composed of small round cells with high nuclear to cytoplasmic ratio.

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**Figure 3.** Neuron-specific enolase immunoreactivity in tumor cells.

weeks after completion of radiotherapy, neurologic examination was normal and postcontrast MRI of the spine and head showed dramatic change in the size of all mass lesions (Figure 6). However, the patient refused to undergo chemotherapy.

### Discussion

Primary neuroectodermal tumors are common tumors in children and are mainly intracranial.<sup>11</sup> These tumors frequently spread throughout the central nervous system via the cerebrospinal fluid and rarely metastasize outside the neuroaxis.<sup>2</sup>

Primary intraspinal primitive neuroectodermal tumors are rare and only 14 cases have been previously reported in the literature.<sup>2-11</sup>

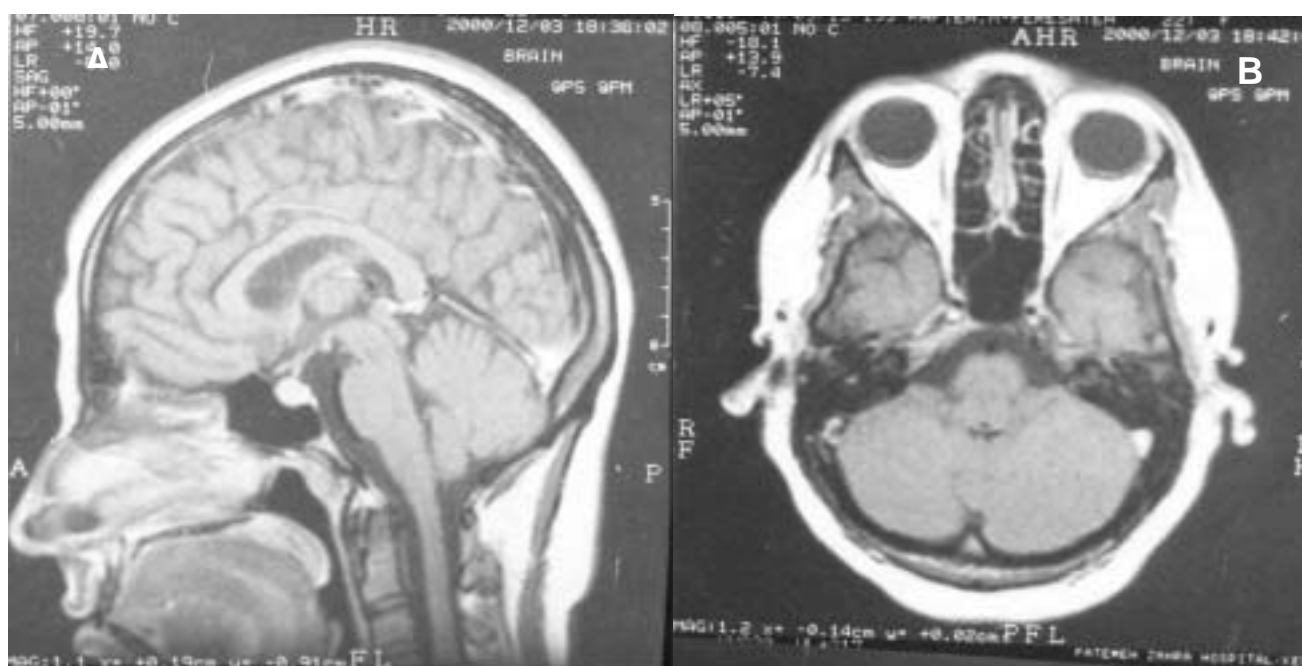
In this case, postcontrast gadolinium-enhanced MRI of the skull failed to reveal any intracranial tumor. The tumor was therefore most likely to be a primitive neuroectodermal tumor. A review of the literature shows that primitive neuroectodermal tumors may arise at any level of the spinal cord and can be intramedullary,<sup>4,11</sup> intradural extramedullary,<sup>5,7</sup> or extradural.<sup>10</sup> These tumors seem to have predilection for the cauda equina, because six of 14 cases show origin from the cauda equina.<sup>2,5,6,8,12</sup> In this case, an intradural extramedullary primitive neuroectodermal tumor was seen at the cervical, thoracic, and thoracolumbar junction.

It has been postulated that PNETs arise from neoplastic transformation of primitive neuroepithelial cells in subependymal zones.<sup>12</sup> Subependymal zones are present in all areas of the central nervous system and may explain the presence of PNETs at locations other than the cerebellum.<sup>2</sup> Spinal PNETs appear to be more common in adults rather than in children in contrast to intracranial PNETs that predominate in children.<sup>2</sup>

There appears to be agreement that, for the management of PNETs, surgery and radiotherapy are essential and chemotherapy is adjuvant.<sup>11</sup> Postoperatively, radiation of the entire cranio-



**Figure 4.** Postcontrast MRI of the spine (A) and head (B) show recurrence of a tumor at original site and at the level of T8-T9, T6 and C1 (arrow).



**Figure 5.** Postcontrast sagittal (A, left) and axial (B, right) MRI of head was failed to reveal any intracranial tumor.

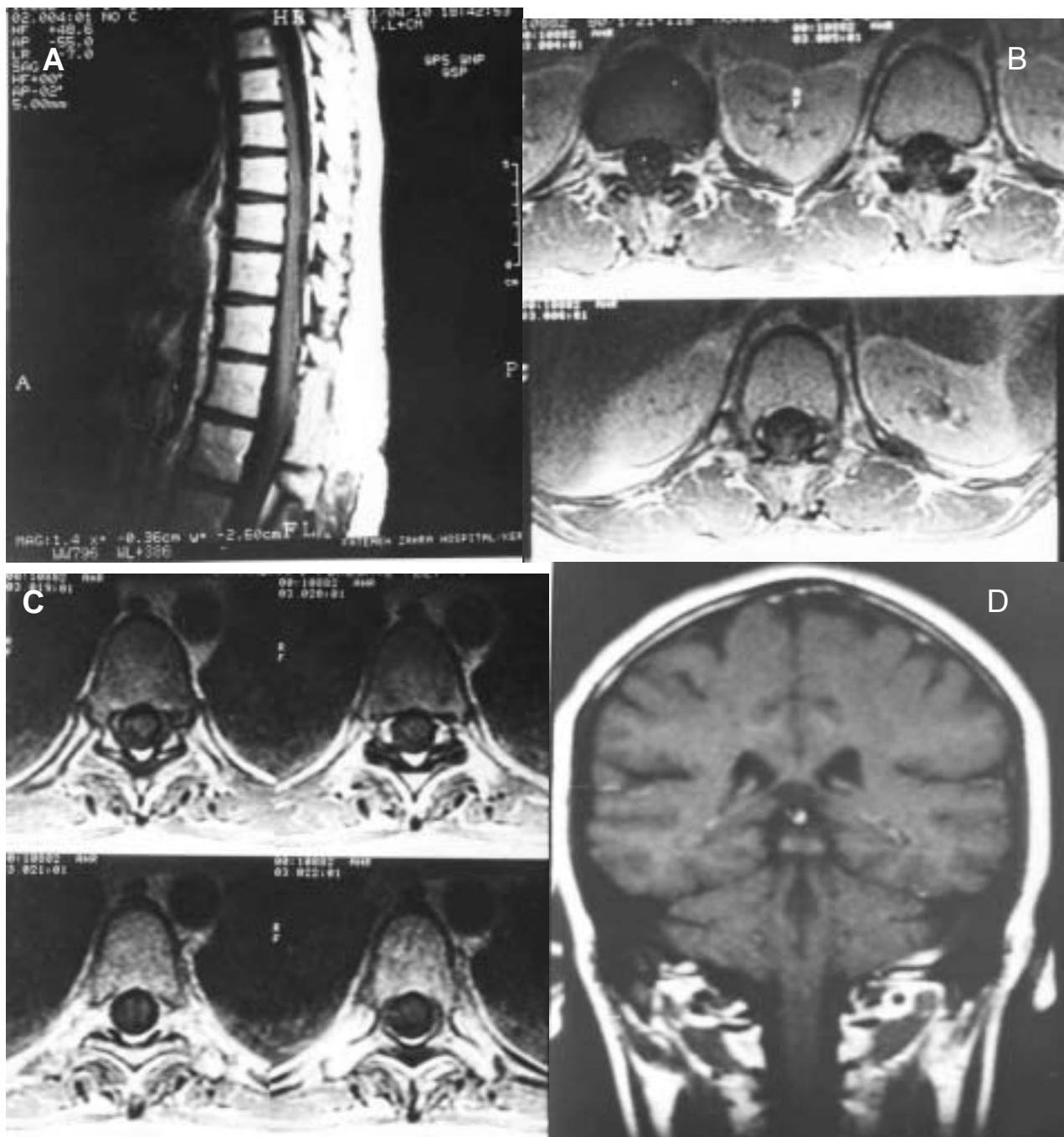
spinal axis is recommended.<sup>13</sup> In this patient, craniospinal radiation was performed and a good initial response to radiotherapy was seen. Because the prognosis for primitive neuroectodermal tumors is usually poor (8 of 14 patients died within 2 years<sup>2</sup>), the patient was scheduled for adjuvant chemotherapy, however, the patient refused and now, 9 months after surgery she is healthy.

The causes of death among these patients include pneumonia,<sup>14</sup> metastatic disease,<sup>7</sup> aggressive local spread of the tumor<sup>8</sup> and progressive spinal cord involvement.<sup>4</sup>

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**Figure 6.** Post contrast MRI of spine and head 8 weeks after completion of radiotherapy. A) sagittal and B) axial at the level of T12 (original site), C) at the level of T8 – T9 and D) coronal view of head and craniovertebral junction. Note the change in tumors size.