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Case Report

Multilevel Primary Intraspinal PNETs in an Infant Associated with Hydrocephalus

Yenidoğanda Hidrosefaliyle Birlikte Görülen Çok Seviyeli Primer Intraspinal PNET

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ABSTRACT

PNETs of the spinal cord are aggressive and local recurrence and/or leptomeningeal spread is common. Primary spinal PNETs are extremely rare and most cases involving the spinal cord are drop metastases from primary intracranial tumors by cerebrospinal fluid. Herewith, we present a 40-day-old infant with multilevel primary spinal PNET at Th12-L1 and L5-S1 levels associated with hydrocephalus occurring nearly 15 days after the operation. According to our knowledge this is probably the first case harboring all these pathologies. Multilevel primary intraspinal PNET in an infant is even rarer and can be associated with hydrocephalus that occurs during the postoperative period.

KEYWORDS: Intraspinal tumor, PNET, Infant, Hydrocephalus

ÖZ

Spinal kordun PNET'leri agresiftir ve lokal rekürens ve/veya leptomeningeal yayılım sık görülür. Primer spinal PNET'ler oldukça nadirdir ve spinal kordu tutan pek çok PNET intrakranial tümörün beyin omurilik sıvısı ile yayılımı sonucu görülür. Biz burada Th12-L1 seviyelerindeki spinal PNET'leri nedeniyle ameliyat edilen ve cerrahiden 15 gün sonra hidrosefali gelişen 40 günlük bir yeni doğanı sunuyoruz. Bizim bilgilerimize göre vakamız tüm bu patolojilere sahip ilk vaka olma özelliğine sahiptir. Çok seviyeli primer intraspinal PNET, spinal kordun PNET'lerine göre çok daha nadir görülür ve cerrahi sonrası dönemde hidrosefali eşlik edebilir.

ANAHTAR SÖZCÜKLER: Intraspinal tümör, PNET, Yenidoğan, Hidrosefali

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INTRODUCTION

Primary intraspinal primitive neuroectodermal tumors (PNETs) are rare and can arise as intramedullary, extramedullary or extradural tumors at any level of the spinal cord (4,17). Primary PNETs of the cauda equina are even less common (7,8,14,12,21). The majority of PNETs that involve the cord or the spinal nerve roots are metastatic in origin and are the result of "drop" metastases from the tumors more superior in the neuroaxis (4,8).

We report a case of primary intraspinal PNET of the conus and left S1 root that presented with progressive paraparesis in a 40-day-old infant.

CASE

A 40-day-old male infant presented at our hospital (Akdeniz University) with a history of progressive lower extremity weakness of 4 days.

Neurological examination revealed bilateral weakness of the lower extremities (2/5 motor strength).

Magnetic resonance imaging (MRI) of the spine revealed an intradural-extramedullar lesion a the L5-S1 level filling the spinal canal and another intradural lesion with conus medullaris invasion at Th12-L1 level (Figure 1A), but his cranial MRI was normal.

The patient was operated on under general anesthesia in the prone position. A T12-L5 laminotomy was performed and dura then opened vertically. At the T12-L1 level the yellowish mass was invading the conus and cauda equina, and at the -S1 level the left S1 root was invaded. Both of the lesions were resected gross totally by microsurgical technique. There was also leptomeningeal infiltration of the mass, mostly at the T12-L1 level.

Postoperative MRI of the spine revealed gross total excision of these lesions (Figure 1B).

Biopsy materials were composed of poorly differentiated round cells disposed in a monotonous sheet or small groups. Tumoral cells had small nuclei and scant cytoplasm (Figure 2). Tumoral cells also showed CD99 and NSE immunoreactivity (Figure 3).

The patient's neurological examination showed improvement following the operation. He was able to move his lower extremities against the gravitational force.

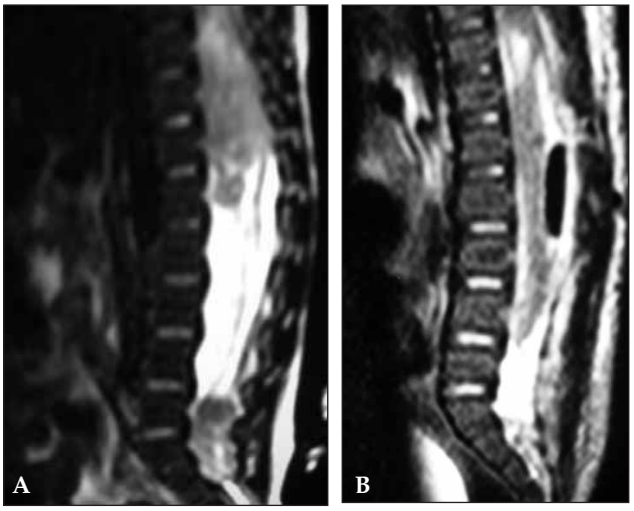

Figure 1: A. *MRI of the spine showing the intradural-extramedullary lesions at Th12-L1 and L5-S1 levels.* ***B.*** *Postoperative spinal MRI with gross total resection of the lesions.*

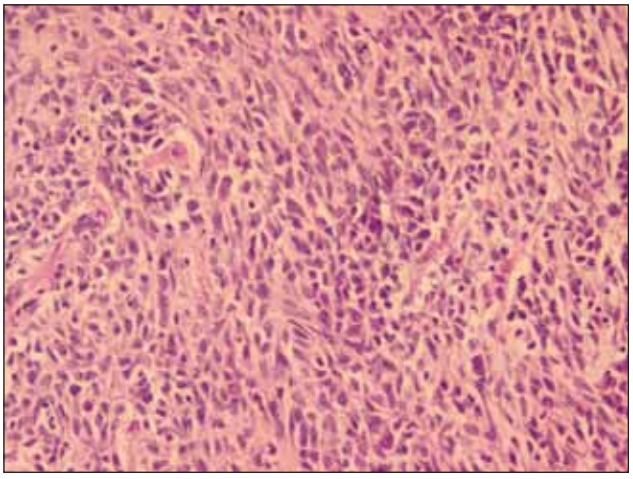

Figure 2: *Tumor consist of small, undifferentiated cells, H&E,X200.*

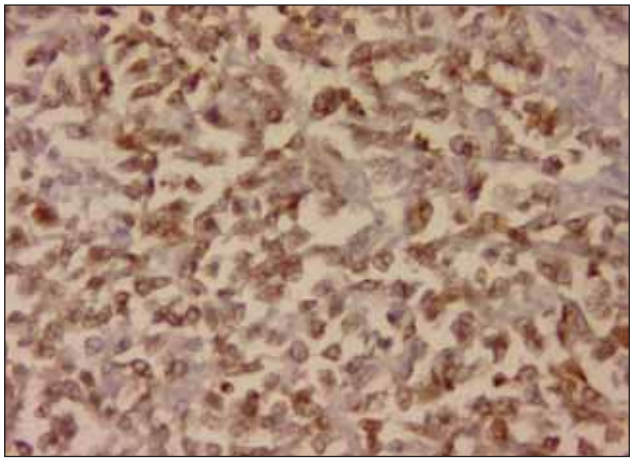

Figure 3: *Tumoral cells showing CD99 and NSE immunoreactivity.*

The patient was given chemotherapy with a combination of vincristine, etoposide, doxorubicine and ifosfamide by the pediatric oncology department as he was under 2 years of age.

Approximately 15 days after the operation he began vomiting and his neurological examination revealed tendency to sleep and slowness of movements. His cranial MRI revealed tetraventricular hydrocephalus. A right ventriculoperitoneal shunt was inserted.

Unfortunately, the patient died due to a pneumonic infection 6 months after the operation.

DISCUSSION

PNETs are malignant small blue neoplasms of children, but can occur at any age. They are composed of predominantly undifferentiated cells and have been traditionally known as medulloblastoma in the cerebellum (2,19).

WHO used the term PNET to describe cerebellar medulloblastomas and other neoplasms that are histopathologically indistinguishable from the latter but that are located at sites in the central nervous system (CNS) other than the cerebellum (9).

Primary spinal PNETs are extremely rare and most cases involving the spinal cord are drop metastases from primary intracranial tumors by cerebrospinal fluid. Intracranial seeding has also been reported in the literature (1,6,10,16). According to our knowledge there are 20 more primary intraspinal PNETs, other than our case, in the literature, but there are only two cases with sacral spinal root involvement (5,22). There is only one case with multilevel spinal PNET and we also believe that our case is also a primary multilevel PNET due to patient's age and tumor size (cited in 22). Furthermore, we could not find any case of a 40-day-old infant harboring primary intraspinal PNET in the literature.

PNETs of the spinal cord are aggressive and local recurrence and/or leptomeningeal spread is common (11). Our case is also probably the first to present with hydrocephalus (HCP). Surgical manipulation and leptomeningeal dissemination could have played a role in the occurrence of HCP (3). The spread of subarachnoid tumor can create an increase in the outflow resistance, explaining the ventricular enlargement and elevated pressure (13,15,18,20).

Although there is no standard therapy for spinal cord or cauda equina PNETs, successful results have been reported using combinations of cyclophosphamide or ifosfamide, cisplatin or carboplatin, and VP-16 (4,12,17).We also preferred chemotherapy because the patient's age was under 2 years, but unfortunately the patient died due to a pneumonic infection after the operation.

CONCLUSION

Primary intraspinal PNET is an extremely rare condition. Multilevel primary intraspinal PNET in an infant is even rarer, and can be associated with hydrocephalus that occurs during the postoperative period.

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