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Olgu Sunumu / Case Report

Primary Spinal Cord Glioblastoma Multiforme Presenting with Transverse Myelitis

Transvers Miyelitle Gelen Bir Primer Spinal Kord Glioblastoma Multiforme Vakası

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ABSTRACT

Primary spinal cord tumors are rarely encountered in childhood period. Ependymomas and pilocytic astrocytomas comprise the majority of spinal cord tumors in children. Spinal glioblastoma multiforme (GM) (grade IV astrocytoma) is a rare clinical entity accounting for only 1-3% of all pediatric intramedullary tumors. We report a 3- year-8- month-old male with primary spinal cord GM who presented with back pain, paraparesis, gait disturbance and loss of sphincter control and initially diagnosed as transverse myelitis.

Key Words: Spinal cord tumors, glioblastoma multiforme, transverse myelitis, childhood

ÖZET

Primer spinal kord tümörlerine çocukluk çağında nadiren rastlanır. Ependimomlar ve pilositik astrositomlar bunların başında gelir. Spinal glioblastoma multiforme pediatrik intramedüller tümörlerin %1-3\'ünü oluşturan nadir bir antitedir.Bu yazıda sırt ağrısı, paraparezi, yürüme bozukluğu ve sfinkter fonksiyon kaybı ile başvuran ve başlangıçta transvers miyelit olarak değerlendirilen 3 yaş 8 aylık erkek hasta sunulmuştur. Klinisyenlere bu şekilde transvers miyelit bulgularıyla başvuran vakalarda intramedüller tümörlerin de ayırıcı tanıda akılda tutulması gerektiği önerilmiştir.

Anahtar Kelimeler: Spinal kord tümörleri, glioblastoma multiforme, transverse myelitis, çocukluk çağı

INTRODUCTION

Primary spinal cord tumors are rarely encountered in childhood period. Intradural intramedullary tumors are responsible for 1-10% of all central nervous system tumors and 30% of spinal tumors in childhood[1-3]. Five percent of intramedullary spinal cord gliomas are encountered in children with neurofibromatosis type 1 (NF1)[4].

Spinal glioblastoma multiforme (GM) is an extremely rare clinical entity that is classified as grade IV astrocytoma by World Health Organization (WHO), accounting for only 1.5% of all spinal cord tumors in children[5]. To our knowledge, there are less than 20 cases of primary spinal cord glioblastoma multiforme in children. Here we report a 3-year-eight-mont-old boy with spinal glioblastoma multiforme with clinical findings resembling transverse myelitis at time of diagnosis.

CASE REPORT

A 3-year-eight-month-old male was admitted to our hospital with symptoms of back pain, weakness in lower limps, disability to walk, enuresis and encopresis having started one week ago. There was no trauma, recent vaccination, infection or a chronic illness in his past medical history. On physical examination, the general status was well with a normal consciousness level. There were multiple café-au-lait spots on the skin with an approximate size of 0.5 cm. The other physical findings were normal. Neurological examination revelaed loss of strength in lower extremity muscles (2/5). Deep tendon reflexes (DTR) and abdominal skin reflexes were absent. Muscle strength was 5/5, and DTRs were present in the upper extremities. There was no pathologic reflexes.

There was no family history of cancer or neurofibromatosis. The full blood count and biochemistry were normal. Cerebral magnetic resonance imaging (MRI) findings were normal. Spinal MRI revealed an intramedullary mass of 130x14 mm, showing irregular contrast enhancement, between the levels of C7-T8 (Figure 1). Laminectomy was performed between T1-T10, and the intramedullary mass was excised with microsurgery. On pathological examination, high grade infiltrative glial tumor (glioblastoma multiforme, WHO Grade IV) was reported (Figure 2). The patient was discharged from the hospital on 13th day of postoperative period with a muscle strength of 2/5, normoactive deep tendon reflexes at lower extremity and intact sphincter control. Radiotherapy and concomittant temozolomide (75 mg/m2) was started one month after surgery. But the treatment was discontiued multiple times because of respiartory tract infections resulting from respiratory complications that developed after surgery. The patient died of a severe lower respiratory tract infection 5 months after the diagnosis.

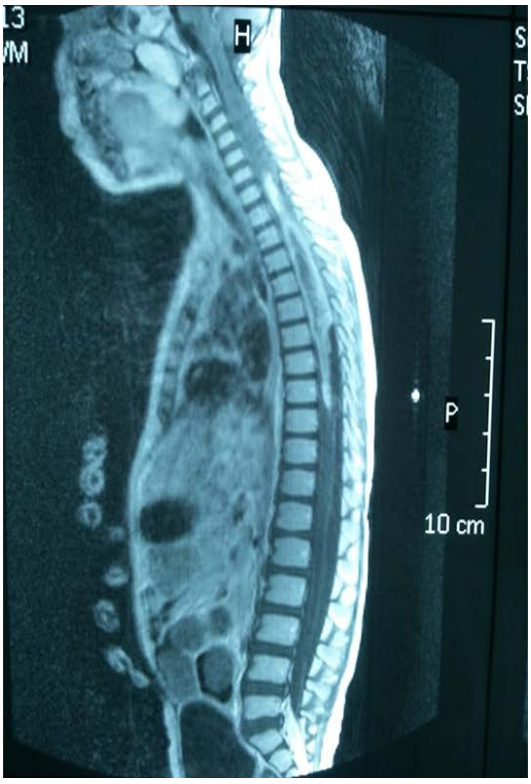

Figure 1. Spinal MRI showing intramedullary with irregular contrast enhancement, between the levels of C7-T8.

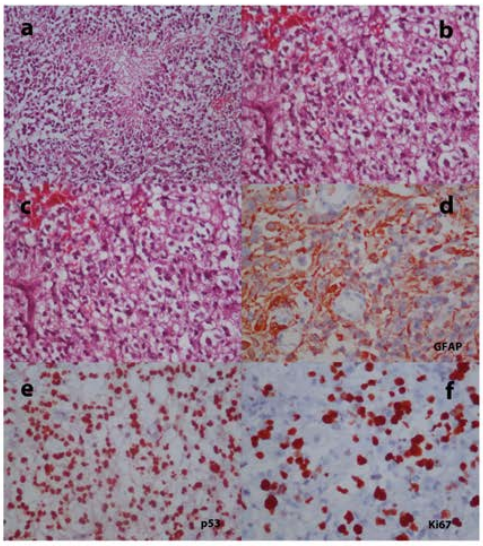

Figure 2.
a) Pseudopalisading necrosis, H-EX200
b) Microvascular-endothelial proliferation, H-EX400
c) Small, round cells dominant area, H-EX400
d) Neoplastic cells are positive for GFAP, X400
e) Nuclear immunoreactivity for p53 protein, X400
f) High proliferative activity, Ki-67X400

DISCUSSION

Primary spinal cord tumors are quite rare in childhood period. Intradural intramedullary tumors are responsible for 1-10% of all central nervous system tumors and 30% of spinal tumors in childhood. Mean age at diagnosis is 10 years. Compared to females, it is 1.3 times more likely to be observed in males[6]. Intramedullary tumors generally seen in children are of gliotic origin. Approximately 60% of them are astrocytomas. Most of them are low grade tumors and they usually originate from thoracic region[1,3,6]. Primary spinal GM is an extremely rare tumor comprising 1-3% of intramedullary spinal cord tumors in children most frequently involving cervical and thoracal regions[7]. Clinical findings include back or neck pain, motor deficits and gait disturbances that were all present in our case.

Neurofibromatosis type 1 is an autosomal dominant neurocutaneous disorder. Diagnostic criteria include six or more café-au-lait spots 1.5 cm or larger in post-pubertal individuals, 0.5 cm or larger in pre-pubertal individuals, two or more neurofibromas of any type or one or more

plexiform neurofibroma, freckling in the axilla or groin, optic glioma, two or more Lisch nodules (benign iris hamartomas), a distinctive bony lesion: dysplasia of the sphenoid bone or dysplasia or thinning of long bone cortex, a first-degree relative with NF1. The diagnosis is based on presence of two or more criteria. Malignancy risk is increased in individuals with NF1. Five percent of intramedullary spinal cord gliomas are seen in children with NF1. The risk of developing an optic gliom is 1000 times, a soft tissue sarcoma is 50 times and a brain or spinal cord tumor is 40 times more than normal population in patients with NF1[8]. Our patient had multiple milimetric café-au-lait spots. However he did not meet the other NF1 diagnostic cirteria. As the diagnosis can be difficult and often delayed in younger patients with no family history, we decided to evaluate our patient for NF1 during follow-up visits.

Spinal GM develops as secondary to metastasis of brain GM in 25% of the cases[9]. On the other hand, the intracranial dissemination of primary spinal GM is extremely uncommon[10]. Dissemination occurs by leptomeningeal involvement rather than intracranial invasion[25,26]. In our case cerebral MRI revealed no brain or meningeal metastasis.

Treatment options include surgery, radiotherapy and chemotherapy. The role of radical surgery in treatment of high grade intramedullary spinal cord tumors is not clear. Conservative approach with tumor biopsy and postoperative radiotherapy rather than agrressive surgery was the recommended management of primary spinal cord GM before the invent of modern neurosurgical techniques using neuroimaging and intra-operative neurophysiology monitoring[7,11,12]. In our patient the intramedullary mass between T1-T10 was resected with microsurgery. Radiotherapy can be given in preoperative or postoperative period. The use of radiotherapy in children under three years is controversial[7]. However, improved survival was reported in an 18-month-old child who received radiotherapy after gross total resection[13]. In our patient, radiotherapy was started after surgical resection but patient died before the completion of radiotherapy due to respiratory complications developed after surgery. The role of chemotherapy is also controversial and a precise data revealing the effectiveness of chemotherapy have not been reported in previous studies[14,15]. Compelling results with concomittant temozolomide and radiotherapy following adjuvant temozolomide were reported in adults with high grade astrocytomas[12]. However, pediatric studies failed to report improved outcomes with temozolomide[16]. Temozolomide was started in our patient concommittantly with radiotherapy. Despite multimodality treatment approaches, prognosis is poor and average life expectancy is 15 months after the diagnosis[5,21,22,23].

In conclusion, being an uncommon condition, the diagnosis of primary spinal cord GM is less frequently considered than infectious or postinfcetios disorders in children with presenting neurological deficits indicating spinal cord involvement. Neuroimaging techniques provide adequate information for differential diagnosis. Due to poor prognosis despite all treatment modalities and rapid progression, prompt management of diagnostic procedures are important. Intramedullary spinal cord tumors should be considered in differential diagnosis of clinical findings resembling transverse myelitis.

Conflict of interests: None

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