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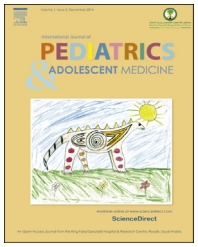

CASE REPORT

Thoracic spinal meningioma in a child with Down syndrome: A case report and review of the literature

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Abstract Down syndrome is the most common genetic chromosomal disorder and occurs in one out of every 700 newborns. It is well-established that individuals with Down syndrome exhibit a unique tumor profile. These individuals are predisposed to certain neoplasms, such as leukemia and other hematological malignancies. However, solid tumors are exceptionally rare. Central nervous system (CNS) tumors in individuals with Down syndrome have been reported in only a small number of case reports. The majority of these tumors are gliomas and germ cell tumors. Meningiomas have yet to be reported in Down syndrome. We report the first case of a meningioma tumor in an individual with Down syndrome. We present a case of spinal meningioma in a 14-year-old boy with Down syndrome.

1. Introduction

Down syndrome is the most common genetic chromosomal disorder and occurs in one out of every 700 newborns [1]. It is well-recognized that individuals with this chromosomal disorder exhibit a unique tumor profile. These individuals have a greater risk of developing hematological malignancies such as acute lymphoblastic and acute megakaryoblastic leukemias. These tumors are 19-fold more common among individuals with Down syndrome than in the

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normal population [2]. Testicular cancer is also markedly more prevalent among individuals with Down syndrome. However, other solid tumors are generally uncommon [3,4]. These observations clearly indicate a role of genetic factors in this altered susceptibility. Such genetic factors remain poorly understood. Tumors of the central nervous system (CNS) in individuals with Down syndrome have been reported in the literature in only a few case reports, and the reported tumors are primarily gliomas and germ cell tumors. Meningiomas have yet to be reported. Here, we present a case of a 14-year-old boy with Down syndrome who was diagnosed with a thoracic spine meningioma. This case report is the first published description of a meningioma in a patient with Down syndrome.

2. Case

A 14-year-old boy known to have trisomy 21 (Down syndrome) based on his morphological features presented with a 2-year history of progressive spastic paraparesis. His weakness progressed in the 4 months before presentation at our institute. He became wheelchair-bound with urinary incontinence. This presentation was initially thought by the referring hospital to be of musculoskeletal origin and related to his genetic disorder. He was overweight with a BMI of 43. He also complained of back pain for the past few months. The patient and his family denied any history of trauma, upper limb symptoms, exposure to ionized radiation, or any family history suggestive of neurofibromatosis disorders or CNS tumors. He went through multiple inpatient and outpatient physiotherapy programs with no clinical improvement, and his symptoms continued to deteriorate with time. On physical examination, the patient displayed a significant mental delay for his age. He was able to communicate with simple dialog only. He had the classical morphological features of Down syndrome in his face, neck, hands, and soles. No obvious cutaneous stigmata suggestive of neurofibromatosis were observed. Cranial nerve examination revealed no obvious abnormalities. An examination of the upper limbs was within normal limits. In his lower limbs, he had bilateral spastic weakness with a motor power of 2—3 on a scale of 5 proximally and 3/5 distally. He had impairment of all sensory modalities in the lower limbs with a partial sensory level at T4. Due to his lower limb weakness, he had been wheelchair-bound for the last 3 months. His muscles tone was increased with brisk deep tendon reflexes, a sustained ankle clonus and an up-going bilateral Babinski's sign. Chromosomal analysis in our institute confirmed the karyotype of trisomy 21;47,XY, +21. Magnetic resonance Imaging (MRI) of the thoracic spine revealed an intradural extramedullary dural-based mass measuring 3 cm in length that was compressing and displacing the spinal cord anteriorly. The lesion extended from the vertebral body level of T5—T6. The lesion was isointense on T1- and T2-weighted images with homogenous enhancement following intravenous gadolinium injection. The lesion also had a small dural-based tail [Fig. 1]. The radiological features were highly suggestive of a spinal meningioma. MRI of the brain and the rest of the spine revealed no other lesions. The patient underwent a surgical intervention that included T5 and T6 bilateral laminectomies, a midline dural incision and resection of the dural-based lesion with coagulation of the tumor bed (Simpson grade II). Intralesional calcifications were noted. The surgery was uneventful with no intraoperative complications. A few days postoperatively, he experienced a cerebrospinal fluid leak that was managed with a temporary lumbar drain. Histopathological study revealed classical transitional meningothelial cells with the psammomatous subtype in many areas [Fig. 2]. His physical evolution was very satisfactory. He exhibited gradual

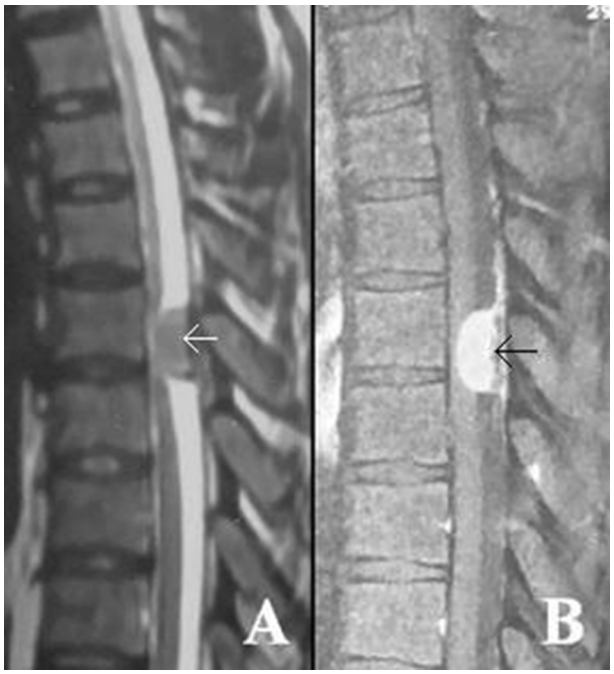

Figure 1 Sagittal MRI showing a dural-based lesion compressing the spinal cord at the level of T5—T6. The mass is isointense on T2-weighted (A) and T1-weighted images with homogenous enhancement following contrast injection (B).

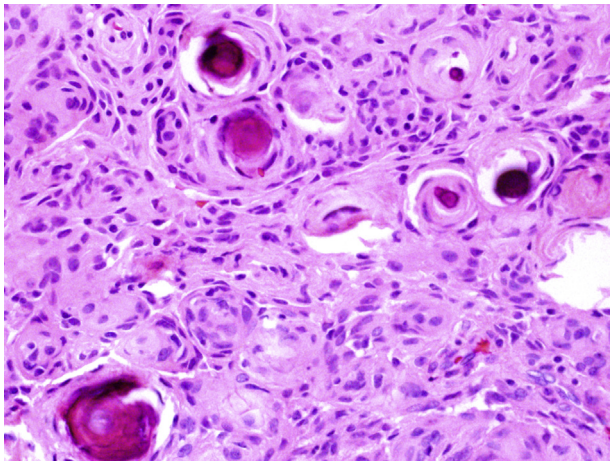

Figure 2 This figure illustrates the arrangement of the cells in a whirling pattern. The cells have round to oval nuclei and exhibit nuclear inclusions in some places. Variable numbers of calcified psammoma bodies were also identified (H&E stain).