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Pediatric Pilocytic Astrocytoma: A Rare Case of Intramedullary Location

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Abstract

Pilocytic astrocytoma is the benign form of astrocytomas. It is most commonly seen in localizations such as the cerebellum, optic nerve, optic chiasm, hypothalamus, third ventricle and peduncle. Its intraspinal localization is much less well known and it represents the rarest location of pilocytic astrocytoma. This tumor is typically well-circumscribed and contrast-enhancing. Currently, only about one hundred cases of intramedullary pilocytic astrocytoma have been published in the world literature. Due to the rarity of this location, most of the literature exists as case reports. We are here reporting our experience with a new case of 11 years-old girl with a spinal pilocytic astrocytoma ranging from the medullary bulbo junction to T4.

Keywords: Pilocytic astrocytoma, Astrocytomas, Spinal tumor, Pediatric tumor

Abbreviations: MRI: Magnetic Resonance Imaging; PA: Pilocytic Astrocytoma

Introduction

Pilocytic astrocytoma (PA) is the benign form of astrocytomas with a long clinical history. It is most commonly seen in localizations such as the cerebellum, optic nerve, optic chiasm, hypothalamus, third ventricle and peduncle. Its intraspinal localization is much less well known and it represents the rarest location of pilocytic astrocytoma. This tumor is typically well-circumscribed and contrast-enhancing. Sixty percent are localized to the cervical and upper thoracic cord and 40% are associated with a tumor cyst or syrinx [1]. Currently, only about one hundred cases of intramedullary pilocytic astrocytoma have been published in the world literature. Due to the rarity of this location, most of the literature exists as case reports. We are here reporting our experience with a new case of 11 years-old girl with a spinal pilocytic astrocytoma.

Case Presentation

A 11-year-old girl patient with no previous medical history presented with progressively deteriorating weakness of the lower extremities, in particular on the left, and urinary and fecal incontinence. Four years before, a progressive "idiopathic" scoliosis was diagnosed with back pain but no imagery was performed; only medical treatment was prescribed. The neurologic examination revealed decreased muscle strength of lower extremities, in particular on the left, and urinary and fecal incontinence. Four years before, a progressive "idiopathic" scoliosis was diagnosed with back pain but no imagery was performed; only medical treatment was prescribed. The neurologic examination revealed decreased muscle strength of lower extremities (spastic paraparesis) with peripheral brachial diplegia with sensitive disorders. Brain magnetic resonance imaging (MRI) and spine MRI were performed. The cranial MRI revealed no pathologic Changes. Spinal MRI revealed an intramedullary lesion ranging from the medullary bulbo junction to T4 with relatively well-circumscribed margins, intermixed cystic and solid parts (Figure 1). The solid portions were moderately hyperintense on T2WI and hypointense on T1WI. The
cystic portions were marked hyperintense on T2WI and hypointense on T1WI. On the contrast-enhanced, MR images showed focal nodule enhancement mainly in peripheral of the solid components of tumor. Antiedema treatment with steroids was prescribed, and then she underwent C1–T4 laminectomy with resection of the spinal cord tumor and drainage of the cysts. Histological examinations showed a WHO Grade I pilocytic astrocytoma. Postoperatively, there is a slight worsening of the motor deficit and an obvious posterior cord syndrome. No adjuvant radiotherapy or chemotherapy received. She was transferred to functional rehabilitation hospital for further care with progressively improvement of motor disorders at 6 months of follow-up.

Figure 1: Cervical-thoracic MRI, Sagittal contrast-enhanced T1-WI (A), sagittal T2-WI (B) and axial T2-WI (C) showing preoperative imaging.

Discussion

Spinal pilocytic astrocytomas occur primarily in the pediatric and adolescent population. It is usually a benign astrocytoma that has excellent prognosis for cure and long term survival, the 5- to 10-year survival for pilocytic astrocytomas (WHO grade 1) has been suggested to be as high as 81% [2]. Pilocytic astrocytomas grow insidiously and symptoms can take months to years to evolve. The symptoms are various depending on the localization of the tumor, the most common are pain, motor and sensorial deficits, and urinary and fecal incontinence, which start occurring on average 2-3 years before diagnosis (The course of the disease ranged from 2 months to 5 years) [3]. We have almost the same symptoms in our case with scoliosis and pain since 4 years.

On imaging, most of Pilocytic Astrocytoma (46.15%) was located in the cervical region, but intramedullary neoplasms affecting the entire spinal cord from the cervicomedullary junction to conus medullaris are also reported in literature with 24 cases [4]. Concerning tumor distribution on axial images, most of PAs (61.5%) were eccentrically located in the spinal cord; PA seems to be more eccentric than ependymoma [5]. The average length of vertebral segments involved were 4.7 segments (range, 1–17 vertebral segments). Eleven vertebral segments were involved in our case.

Usually, these tumors had relatively well-circumscribed margins and their consistency is intermixed cystic and solid. Syringomyelia may be associated. In general, the solid portions of tumors are moderately hyperintense on T2WI and hypointense on T1WI but the cystic portion is hyperintense on T2WI and hypointense on T1WI. On the contrast-enhanced MR images, most of reported cases (53.8%) showed focal nodule enhancement (intense or mild) of the solid components of tumor [5], this enhancement is smaller than ependymoma. Our imaging results are similar to the literature.

The management of these tumors is still controversial. In the past such tumors were biopsied and then treated by radiation therapy. Nowadays, Surgery is considered first-line therapy, because it is associated with long-term survival [6]. Although, total removal of these tumors are considered difficult and aggressive surgery increases the risk of neurologic morbidity [7]. The potential to recover lost neurological function is very limited. Thus, a good preoperative neurological status is the main predictor for a satisfying postoperative outcome.

The need for adjuvant therapy depends on the extent of resection. The role of radiotherapy is still under debate. Minehan et al. [2], reported a higher survival rate with
postoperative radiotherapy, but this was more significant for fibrillary astrocytomas than for pilocytic astrocytomas. However tolerance of the spinal cord to radiotherapy is limited and there is risk of radiation injury. Colnat-Coulbois et al. [8] proposed an interstitial intracavitary rhenium 186 brachytherapy in managing of recurrent spinal cord cystic astrocytomas. This treatment achieved excellent stabilization of the cyst with minor side effects and dramatic improvement of neurologic deficits.

Conclusion

Intramedullary astrocytomas are rare tumors. The clinical symptomatology is nonspecific. Spinal MRI is quite evocative, showing an iso or slightly hypo-intense lesion in T1, hyper-intense in T2 and often associated with cysts. The most complete surgical excision possible is necessary to ensure a good evolution of the patients.

Declarations

The authors declare that there are no conflicts of interest regarding the publication of this article.

References


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