Case Report

Spinal meningiomas in pediatric patients – A case series and literature review

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INTRODUCTION

Meningiomas are the most frequent intracranial tumors in adults accounting for 30% of all intracranial neoplasms. On the contrary, meningiomas comprise only 0.4–4.6% of the central nervous system tumors in pediatric patients. The number of meningiomas increases with age. Pediatric lesions are characterized by an intense progression of the disease, higher recurrence ratio, and more aggressive biological behavior. Patients with diagnosed neurofibromatosis type 2 (NF2) should be included in the dedicated diagnostic and therapeutic programs coordinated by phacomatosis centers. Meningiomas are most often multiple in patients with NF2. In those patients, meningiomas may predispose to worse prognosis and higher mortality. Therefore, patient with NF2 requires special attention. Spinal meningiomas constitute about 5.9–15% of meningiomas in pediatric patients. Those tumors are mostly benign lesions.
Back pain is usually the first symptom of growing tumors. Unfortunately, patients are usually admitted to hospital too late; with neurological deficits and gait disturbances.[14,16]

MATERIALS AND METHODS

Between the years 2000 and 2017, three patients below 18 years of age with spinal meningiomas were hospitalized and operated in the Pediatric Neurosurgery Department. The study group consisted of two girls and one boy. Two patients were diagnosed with NF2. The diagnosis of NF2 was based on the Manchester Clinical Diagnostic Criteria. Gene mutation research was not a part of the protocol. Data were obtained retrospectively from medical histories. Consent was granted by each patient and patient’s parents. Literature review was based on the data collected from the National Center of Biotechnology Information.

CLINICAL PRESENTATION

Case report 1

A 17-year-old boy was hospitalized for the diagnostic management of progressive focal neurological deficits. In clinical examination, he presented micro and retrognathia. In neurological examination, the patient presented right-side hemiparesis, impaired gait, and disturbances of superficial and deep sensation. When his medical history was taken, the patient reported past reconstruction of the temporomandibular joint. Magnetic resonance imaging (MRI) showed a tumor size 23 × 15 × 8 mm adhering to the posterior-lateral surface of the spinal cord at the C1/C2 level. The pathological mass showed strong enhancement after intravenous contrast medium administration. The tumor caused compression and displacement of the spinal cord. In the center of the spinal cord, the tumor was located at the C2 level [Figure 1]. A unilateral spinal nerve schwannoma 5 mm in diameter was located at the C4 level. MRI also showed bilateral vestibular schwannomas. Resection of the cervical meningioma with a partial resection of the C1 and C2 vertebrae was performed. The histopathology result was meningioma psammomatosum. The diagnosis of NF2 was based on the Manchester Clinical Diagnostic Criteria. After the surgery, gait improvement was observed. Cause of legal age follow-up was limited to 1 year.

Case report 2

A 10-year-old girl with muscle weakness involving the lower limbs and unstable gait was hospitalized in the Neurosurgery Department. The neurological examination demonstrated spastic paraparesis of the lower limbs, bilateral plantar reflex, and lack of skin abdominal reflexes. The abnormalities presented on the left side included: clonus and foot drop, exaggerated ankle reflex, weakness of the knee reflex, and positive pronator drift. The patient presented normal superficial and deep sensation. The physical examination revealed muscular atrophy of the left calf. Thoracic MRI showed a small tumor 7 × 5 × 8 mm in size at the Th11 level [Figure 2a]. Hemilaminectomy with total tumor resection was performed. After the surgery, the patient presented progression of lower limbs paraparesis. Control cervical and thoracic MRI demonstrated a massive tumor 24 × 11 × 15.5 mm in size at the Th3/Th4 level [Figure 2b]. The tumor significantly compressed and displaced the spinal cord. After total gross tumor resection, regression of lower limbs paresis was observed [Figure 2c]. Two weeks after surgery, muscle strength of the lower limbs was rated as 3 points in the Lovett’s scale. In follow-up, limb paresis has tendency to regression. The histopathology result was meningioma psammomatosum. Three years later, the girl was hospitalized due to back pain and progression of gait disturbance. Control thoracic MRI showed multiple meningiomas at the Th2 to L1 level [Figure 3a]. Cervical MRI demonstrated a pathological mass at C2–C3 and at C6–C7 level with spinal

Figure 1: Preoperative magnetic resonance imaging (MRI) scans of Patient No. 1 with neurofibromatosis type 2: (a) Axial T2-weighted MRI with contrast shows a meningioma 23 × 15 × 8 mm in size at the C1–2 level. The tumor adheres to the posterior-lateral side of the spinal canal. The meningioma presses and displaces the spinal cord. (b) Sagittal T2-weighted MRI with contrast shows the meningioma and coexisting intraspinal ependymoma sized 11 × 9 × 5 mm at the C2 level. (c) Coronal T1-weighted MRI with contrast shows the meningioma before surgery.
corticospinal compression [Figure 3b]. Schwannomas of the spinal nerves were situated at the levels from C5 to C7. Bilateral vestibular schwannomas were also detected on the MRI scan [Figure 3c]. A right-side hemilaminectomy at the C5–C7 levels and tumor resection was performed. Three months after surgery, spastic muscle tension significantly regressed, and the lower extermities paresis was minimal. The patient was diagnosed as NF2 based on the Manchester Clinical Diagnostic Criteria.

Case report 3

A 14-year-old girl was admitted to hospital because of deterioration of the lower limbs paraparesis and back pain. The patient reported a 2-year history of symptom progression. A MRI scan showed a tumor at the Th4 level compressing the spinal cord. A laminectomy at the Th3-Th4 levels with tumor resection was performed. The histopathology result was menigioma psammomatosum. The symptoms gradually receded after the surgery. Three months after surgery, muscle strength of the lower limbs was rated as 4–5 points in the Lovett’s scale. In 2-year follow-up patients did not present further neurological deterioration. A control MRI scan did not show tumor recurrence or new tumors.

DISCUSSION

Pediatric meningiomas specification

Pediatric meningiomas are characterized by specific biological features including tumor histology, recurrence ratio, location, and prognosis as compared to tumors observed in the adult population. In children, meningiomas are extremely rare, representing 0.4–4.6% of CNS tumors. Meningiomas are more common in older children and adolescents. In infants, meningiomas are extremely rare. Only single cases were described in the literature. Pediatric meningiomas are more frequent in males. A high-grade type meningiomas are more frequent in girls. Females are diagnosed at an earlier age. In the pediatric group the possibility of association...
of genetic syndromes have been taken into consideration. Up to 53% of pediatric meningiomas are associated with NF2, and the risk of NF2 in children with meningioma is estimated as approximately 20%.\[^{10}\] Thujijs et al. suggested that multiple meningiomas could be the first manifestation of NF2.\[^{14}\] Meningiomas in young patients are characterized by a more aggressive biological behavior, tendency to rapid grow, are likely to recur after a short latency period, and have propensity to malignancy.\[^{7,15,17,22}\] Those biological features determine the overall poor prognosis.\[^{17}\]

**Histology**

Pediatric meningiomas are a heterogeneous group of tumors. In children with NF2, meningiomas usually present atypical histological features, such as the papillary variant or clear cell type.\[^{11}\] Kotecha et al. reported high-grade meningiomas (WHO II and III) in 7.2% of pediatric patients as compared to only 1–2.8% of adults.\[^{11}\] Meningiomas appear in the spine only in 5.9–15% of cases. The cervical and thoracic regions are especially typical for spinal meningiomas.\[^{14,20,22}\]

The most common histological types of spinal tumors include psammomatous, meningothelial, and transitional meningiomas.\[^{14,16}\] In genetic syndromes such as NF2, the neoplasms are not as aggressive as the sporadic ones.\[^{11,16}\]

**High-grade tumors**

At present, there is no effective treatment for pediatric patients with high-grade meningiomas. An alternative treatment of inaccessible tumors and histologically aggressive neoplasm is stereotactic radiosurgery (SRS). These tumors are rare and require special methods of treatment.\[^{12,11}\] The patients require frequent and systematic follow-up examinations.

**Symptoms**

Symptoms are mainly the result of spinal cord compression.\[^{14}\] Frequently, back pain is the first symptom of the growing tumor mass. Subsequently, neurological deficits appear, such as sensor deficits, limbs weakness, and gait disturbances.\[^{14,16,20}\]

**Therapeutic strategy and treatment limitations**

Patients with asymptomatic small benign meningiomas can be observed, but in symptomatic patients, complete surgical resection should be performed.\[^{5}\] A chance to perform total gross resection is an important prognostic factor. Surgery is often curative, but does not eliminate the risk of relapse.\[^{4,11}\] In young patients, the second-look surgery is better than radiotherapy (RTH). RTH should be recommended only in recurrences or high-grade tumors, and in surgically inaccessible locations.\[^{10,13,21}\] Children with Grade II meningiomas can be radiated when they reach the age of 8 years of life. In cases of the WHO Grade III meningiomas, fractioned RTH is recommended for children older than 3 years.\[^{8,13}\] Horiba et al. reported a possibility of SRS in a 2-year-old patient.\[^{6}\] Kondziolka et al. reported successful SRS treatment of meningiomas.\[^{8}\] High-grade meningiomas tend to recur after total surgical resection and RTH.\[^{13}\] Minniti et al. reported a high intracranial tumor control rate after SRS in the range of 85–97% at 5-year follow-up.\[^{12}\] Notwithstanding, Dirks et al. proved that NF2-related meningiomas were not as sensitive to SRS as sporadic tumors.\[^{10}\] The use of RTH and SRS is still limited due to the possibility of long-term late toxic effects, such as intellectual development impairment, focal, and neurological deficits.\[^{8}\] In each case, risk and benefits of RTH and SRS in pediatric patients should be take under consideration.\[^{13}\] At present, no sufficient volume of research addressing RTH and SRS in the pediatric group of spinal meningiomas is undertaken.\[^{1,22}\]

**Follow-up**

Patients with NF2 are at a high risk of developing progressive and recurrent tumors that should be carefully monitored. Dirks et al. suggested a long postoperative follow-up period.\[^{5}\] For all cases, surgical treatment is the “treatment of choice.” The extent of resection is an important factor affecting tumor recurrence.\[^{10}\] Nevertheless, spine cord protection should be a priority.\[^{22}\]

**CONCLUSION**

Summarizing, spinal meningiomas are rare tumors in the pediatric population. In children, they often are the first manifestation of NF2. Therefore, the affected child should be under strict control and a MRI-scan should be periodically performed. Patients with asymptomatic small benign meningiomas can be observed, but in symptomatic patients, complete surgical resection should be performed. Adjuvant RTH in children should be recommended only for selected cases.

**Author contributions**

Paula Piątek: Substantial contribution to conception and design, acquisition of data, or analysis and interpretation of data; final approval of the version to be published; agreement to be accountable for all aspects of the work thereby ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved, drafting the article or revising it critically for important intellectual content.

Stanisław Kwiatkowski: substantial contribution to conception and design, acquisition of data, or analysis and interpretation of data; drafting the article or revising it critically for important intellectual content.
Olga Milczarek: senior author, substantial contribution to conception and design, acquisition of data, or analysis and interpretation of data; drafting the article or revising it critically for important intellectual content.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Antinheimo J, Haapasalo H, Halite M, Tatagiba M, Thomas S, Brandis A, et al. Proliferation potential and histological features in neurofibromatosis 2-associated and sporadic meningiomas. J Neurosurg 1997;87:610-4.
2. Blakeley JO, Evans DG, Adler J, Brackmann D, Chen R, Ferner RE, et al. Consensus recommendations for current treatments and accelerating clinical trials for patients with neurofibromatosis Type 2. Am J Med Genet A 2012;158A:24-41.
3. CBTRUS. CBTRUS Statistical Report: Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2004-2006. Hinsdale: Central Brain Tumor Registry of the United States; 2010.
4. Chamberlain MC, Blumenthal DT. Intracranial meningiomas: Diagnosis and treatment. Expert Rev Neurother 2004;4:641-8.
5. Dirks MS, Butman JA, Kim HJ, Wu T, Morgan K, Tran AP, et al. Long-term natural history of neurofibromatosis Type 2-associated intracranial tumours. J Neurosurg 2012;117:109-17.
6. Goutagny S, Kalamardies M. Meningiomas and neurofibromatosis. J Neurooncol 2010;99:341-7.
7. Grossbach AJ, Mahaney KB, Menezes AH. Pediatric meningiomas: 65-year experience at a single institution. J Neurosurg Pediatr 2017;20:42-50.
8. Horiba A, Hayashi M, Tamura N, Chiba K, Aihara Y, Kawamata T. Gamma knife treatment of malignant infantile brain tumors-case report. J Radiosurg SBRT 2018;5:249-53.
9. Kondziolka D, Madhok R, Lunsford LD, Mathieu D, Martin JJ, Niranjan A, et al. Stereotactic radiosurgery for convexity meningiomas. J Neurosurg 2009;111:458-63.
10. Kotecha RS, Junckerstorff RC, Lee S, Cole CH, Gottardo NG. Pediatric 406 meningioma: Current approaches and future direction. J Neurooncol 2011;407:1-10.
11. Kotecha RS, Pascoe EM, Rushing EJ, Rorke-Adams LB, Zwerdling T, Gao X, et al. Meningiomas in children and adolescents: A meta-analysis of individual patient data. Lancet Oncol 2011;12:1229-39.
12. Minniti G, Amichetti M, Enrici RM. Radiotherapy and radiosurgery for benign skull base meningiomas. Radiat Oncol 2009;4:42.
13. Norden AD, Drappatz J, Wen PY. Advances in meningioma therapy. Curr Neurol Neurosci Rep 2009;9:231-40.
14. Sandalciglu IE, Hunold A, Müller O, Bassiouni H, Stolke D, Asgari S. Spinal meningiomas: Critical review of 131 surgically treated patients. Eur Spine J 2008;17:1035-41.
15. Santos MV, Furlanetti L, Valera ET, Brassesco MS, Tone LG, De Oliveira RS. Pediatric meningiomas: A single centre experience with 15 consecutive cases and review of literature. Child Nerv Syst 2012;28:1887-96.
16. Setzer M, Vatter H, Marquardt G, Seifert V, Vrionis FD. Management of spinal meningiomas: Surgical results and a review of the literature. Neurosurg Focus 2007;23:E14.
17. Sheikh BY, Siqueira E, Dayel F. Meningioma in children: A report of nine cases and review of the literature. Surg Neurol 1996;45:328-35.
18. Thuijs NB, Uitdehaag BM, van Ouwerkerk WJ, van der Valk P, Vandertop WP, Peerdeman SM. Pediatric meningiomas in the Netherlands 1947-2010: A descriptive epidemiological case study. Childs Nerv Syst 2012;28:1009-15.
19. Traunecker H, Mallucci C, Grundy R, Pizer B, Saran F, Children’s Cancer and Leukaemia Group. Children’s Cancer and Leukaemia Group (CCLG): Guidelines for the management of intracranial meningioma in children and young people. Br J Neurosurg 2008;22:13-25; discussion 24-5.
20. Wang XQ, Zeng XW, Zhang BY, Dou YF, Wu JS, Jiang CC, et al. Spinal meningioma in childhood: Clinical features and treatment. Child Nerv Syst 2012;28:129-36.
21. Wen PY, Quant E, Drappatz J, Beroukhim R, Norden AD. Medical therapies for meningiomas. J Neurooncol 2010;99:365-78.
22. Wu L, Yang Ch, Liu T, Fang J, Yang J, Xu Y. Clinical features and long-term outcomes of pediatric spinal meningiomas. J Neurooncol 2017;133:347-55.

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