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Case Report

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

Paula Piątek[1], Stanisław Kwiatkowski[2] Olga Milczarek[2]

[1]Department of Neurosurgery, St Lukas Hospital in Tarnów, Poland, [2]Department of Childrens' Neurosurgery, Faculty of Medicine, Jagiellonian University Medical College, Kraków, Poland.

E-mail: *Paula Piątek - paulapiatek89@gmail.com; Stanisław Kwiatkowski - stkwiatkowski@o2.pl; Olga Milczarek - olguniam@wp.pl

***Corresponding author:**
Paula Piątek,
Department of Neurosurgery,
St Lukas Hospital in Tarnów,
Poland.

paulapiatek89@gmail.com

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ABSTRACT

Background: Meningiomas are the most frequent intracranial tumors in the adult population; however, they are rare in pediatric patients. In children, meningiomas often require further diagnosis of genetic comorbidities. As many as, 50% of young patients with meningiomas suffer from neurofibromatosis type 2 (NF2). Spinal meningiomas include only 10% of pediatric meningiomas.

Case Description: Between 2000 and 2017, three children were hospitalized in the Neurosurgery Department. The patients reported prolonged periods of increasing neurological symptoms. In each case, a total gross tumor resection was performed. Histopathology result in each patient was meningioma psammomatosum. Only one girl required adjuvant radiotherapy (RTH) due to recurrent tumors. Magnetic resonance imaging (MRI) showed spinal nerves schwannomas and bilateral vestibular schwannomas in two patients with NF2.

Conclusion: A slow tumor growth is characteristic of spinal meningiomas. Back pain is a frequent initial symptom of a slowly growing tumor mass. Subsequently, neurological deficits gradually increase. Patients require a long follow-up period and control MRI-scan. Children with diagnosed spinal meningioma should be strictly controlled because of the high risk of their developing other tumors associated with NF2. Surgical resection is the primary treatment modality of meningiomas. Adjuvant RTH should be recommended only for selected patients.

Keywords: Neurofibromatosis type 2, Pediatric meningiomas, Spinal meningiomas

INTRODUCTION

Meningiomas are the most frequent intracranial tumors in adults accounting for 30% of all intracranial neoplasms.[3] On the contrary, meningiomas comprise only 0.4–4.6% of the central nervous system tumors in pediatric patients.[10] The number of meningiomas increases with age.[18] Pediatric lesions are characterized by an intense progression of the disease, higher recurrence ratio, and more aggressive biological behavior.[18] 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 NF 2.[6] In those patients, meningiomas may predispose to worse prognosis and higher mortality. Therefore, patient with NF2 requires special attention.[2] Spinal meningiomas constitute about 5.9–15% of meningiomas in pediatric patients.[18,19,22] 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

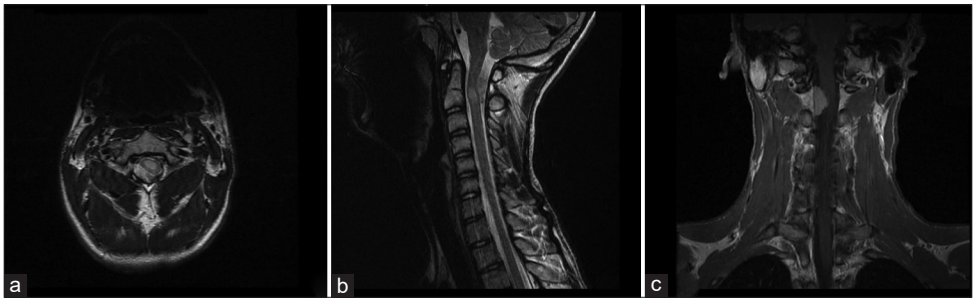

Figure 1: Preoperative magnetic resonance imaging (MRI) scans of Patient No. 1 with neurofibromatosis type 2: (a) Axial T2-weigthed MRI with contrast shows a meningioma 23 × 15 × 8 mm in size at the C1–2 level. The tumor adheres to the posterior-lateral side of the spinal canal. The meningioma presses and displaces the spinal cord. (b) Sagittal T2-weigthed MRI with contrast shows the meningioma and coexisting intraspinal ependymoma sized 11 × 9 × 5 mm at the C2 level. (c) Coronal T1-weigthed MRI with contrast shows the meningioma before surgery.

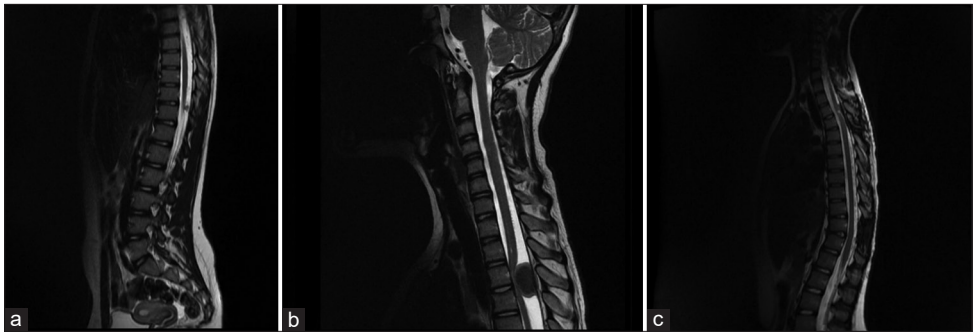

Figure 2: Pre- and post-operative imaging of patient No. 2: (a) Pre-operative sagittal T2-weigthed thoracic MRI of a 10-year-old girl. A small meningioma 7 × 5 × 8 mm at the Th11 level. (b) Sagittal T2-weigthed cervicothoracic junction MRI exposes a massive meningioma 24 × 11 × 15.5 mm at the Th3-Th4 level. The tumor presses and displaces the spinal cord. (c) Post-operative control sagittal T2-weigthed cervical and thoracic MRI. A post-operative reaction at the Th11 level. At the Th3/Th4 level, MRI shows a postoperative reaction or tumor residual mass. A MRI control scan was performed 6 months after the primary surgery.

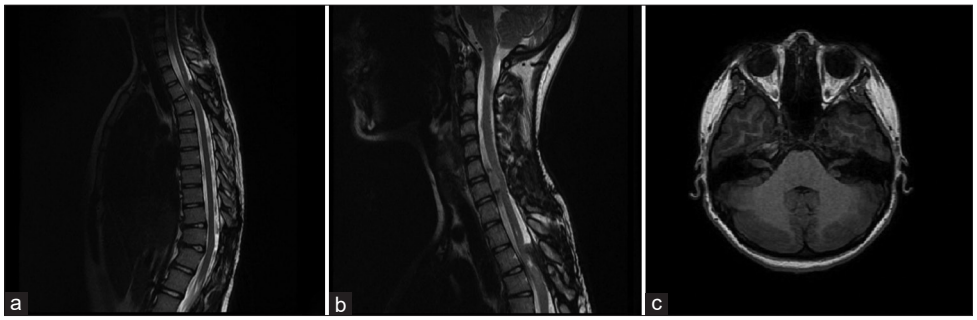

Figure 3: MRI scan performed after 3-year follow-up of Patient No. 2: (a) Sagittal T2-weigthed thoracic MRI shows multiple small meningiomas at the Th2, Th5, Th8, Th9, Th11, Th12/L1 levels, and a tumor at the Th3-Th4 level sized 17 × 7 × 27 mm causing spinal cord compression. (b) Sagittal T2-weigthed cervical MRI presents tumors at the C2–3 and C6–7 levels with spinal cord compression. (c) Axial T2-weighed cervical MRI shows bilateral vestibular schwannomas.

cord 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 month after surgery, spastic muscle tension significantly regresses, 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 meningioma 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.[3] In children, meningiomas are extremely rare, representing 0.4–4.6% of CNS tumors.[10] Meningiomas are more common in older children and adolescents.[7,20] In infants, meningiomas are extremely rare. Only single cases were described in the literature.[7] Pediatric meningiomas are more frequent in males.[20,22] A high-grade type meningiomas are more frequent in girls. Females are diagnosed at an earlier age.[7] 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%.[15] Thuijs *et al*. suggested that multiple meningiomas could be the first manifestation of NF2.[18] 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.[1,10]

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.[2,11] 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.[2,13,21] 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.[8] Kondziolka *et al*. reported successful SRS treatment of meningiomas.[9] 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.[5] 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.[2] At present, no sufficient volume of research addressing RTH and SRS in the pediatric group of spinal meningiomas is undertaken.[2,22]

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.[20] 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.

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