Natural history of primary paediatric optic nerve sheath meningioma: case series and review

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ABSTRACT

Purpose To study the natural history, clinical and radiological characteristics of primary paediatric optic nerve sheath meningioma (PPONS).

Methods Retrospective study of eight paediatric patients who were treated between 1994 and 2016 at the University Hospital Zurich, Switzerland and the Royal Adelaide Hospital, Australia. Clinical records and imaging studies were reviewed.

Results The mean age at presentation was 11 years (range: 6–17 years). There were six female patients and two male patients, 2/8 patients had associated neurofibromatosis type 2. Patients were followed up for 71–297 months (mean 156±70 months). 6/8 patients were observed through the course of their disease and 2/8 patients were treated with radiotherapy. 2/8 patients who were observed had minimal change in vision and did not experience tumour growth after long-term follow-up.

Conclusions This is the largest PPONS case series with long-term data on patients treated conservatively. We highlight that a small subset of these tumours are indolent and can be managed using observation alone.

INTRODUCTION

Optic nerve sheath meningioma (ONSM) is a proliferation of meningothelial cells within the nerve sheath of the orbital or intracranial optic nerve.1 ONSM is rare in persons younger than 20 years of age. ONSM may either be a primary tumour of the optic nerve arachnoid or a secondary tumour due to orbital extension from an intracranial site.

Neurofibromatosis type 2 (NF2) is an autosomal dominant syndrome with an estimated incidence of 1 in 33 000–40 000.2,3 The disease is related to inactivation of a tumour suppressor gene on chromosome 22q12, which gives rise to the development of sporadic and NF2-associated tumours of the central nervous system (CNS), in particular schwannomas and meningiomas. While ONSM account for 1%–2% of all meningiomas, these tumours occur in up to 44% of patients with NF2, particularly if the disease is childhood onset.2

Surgical resection was commonly performed in early reports primary paediatric ONSM (PPONS). However, in most cases surgery offered little benefit and accelerated visual deterioration. The recent trend has been towards primary irradiation using stereotactic, three-dimensional, conformal and fractionated techniques.4–7 There has been one reported case of PPONS managed using observation alone.8

There is little data on the long-term efficacy and safety of radiotherapy for PPONS. The limited use of radiotherapy has been advocated to minimise complications, particularly to the developing cortex and hypothalamic–pituitary system.9

Patients with NF2 carry an additional potential risk of radiation-induced secondary tumours owing to their tumour suppressor gene defect. Further data on the clinical and radiological progression of PPONS will help to counsel patients on the risks and benefits of treatment versus conservative observation.

MATERIALS AND METHODS

This study was conducted in accordance with the Declaration of Helsinki. We conducted a retrospective chart review of eight patients less than 20 years of age who were diagnosed with PPONS at the Royal Adelaide Hospital, Australia (n=4) and the University Hospital Zurich, Switzerland (n=4) between 1994 and 2017. The diagnosis of PPONS was made on the basis of characteristic clinical and radiographic findings.10–13 Cases were excluded when the diagnosis was uncertain.

The following patient characteristics were recorded: age at presentation, sex, affected side, concomitant diagnosis of NF2 as per the Manchester Group criteria,14 15 clinical features including visual acuity, proptosis, pain, limitation of extraocular movements, optic disc swelling or atrophy and presence of optociliary shunt vessels.

Modality of radiological confirmation was recorded. Radiological features including tumour size, morphological configuration, borders, presence of calcification and tumour extension were noted, as were follow-up period, final visual acuity and any deaths. Data are presented as mean values±SD and as percentages.

For our literature review, we conducted a MEDLINE search of the English-language medical literature for the period between 1970 and 2017 using the following keywords: optic nerve sheath meningioma, orbital meningioma, optic nerve tumor, primary, children and paediatric.

RESULTS

Patient characteristics and association with NF2

Results are summarised in table 1. There were six female and two male patients. The mean and median age at presentation was 11 years (range: 6–17 years). Four patients had left-sided tumours, three had right-sided tumours, one had bilateral tumours. Two patients met the Manchester Group diagnostic...
criteria for NF2. Patient 6 was a 13-year-old girl who presented with a right PPONSM. This patient also had multiple intracranial and spinal meningiomas, neuromas of bilateral III, VIII, IX and X cranial nerves and a meningioma of the left XII cranial nerve. Patient 8 was a 9-year-old girl who presented with a left PPONSM. This patient also had bilateral acoustic neuromas and a conus medullaris ependymoma. Patient 7 was a 13-year-old boy who presented with a right PPONSM and had another intracranial meningioma. This patient had one second-degree relative who was suspected of having NF2; however, they did not meet the diagnostic criteria for NF2. Patient 2 presented with bilateral PPONSM and did not have NF2 or any other intracranial pathology.

Clinical features

Decreased vision was the most common presenting complaint in 7/9 eyes. Four out of nine eyes (44%) had a visual acuity of 20/40 or better on initial examination, 1/9 (11%) eyes had vision between 20/60 and 20/200. Three out of nine eyes (33%) had less than 20/200 vision. Six out of nine eyes had limitation of movement and 4/9 eyes had proptosis. Optic disc changes were observed in 6/9 eyes. Two out of nine eyes had optic disc atrophy and 4/9 eyes had optic disc swelling (one of these eyes later developed optic atrophy). Optociliary shunt vessels were observed in 1/9 eyes. Onset of symptoms was chronic (months) in 5/9 cases and subacute (weeks) in 3/9 cases.

Radiological features

MRI and CT were performed during first presentation. MRI was used for all follow-up scans. Tumour configuration was fusiform in 6/9 cases, tubular in the bilateral case and globular in 1/9 cases. Eight out of nine tumours had well-defined margins and 1/9 had irregular margins. Four out of nine tumours had associated calcification. Four out of nine tumours involved the entire length of the intraorbital nerve, 4/9 tumours involved the posterior segment of the intraorbital optic nerve and 1/9 tumours involved the anterior half of the nerve. Intracranial extension was seen in 2/9 tumours on initial presentation. In both of these cases the tumour extended to the prechiasmal region.

Treatment and complications

Final follow-up occurred between 71 and 297 months after initial presentation (mean 156±70 months). Six out of eight patients were observed through the course of their disease and 2/8 patients were treated with radiotherapy using external beam radiation. Radiological growth was recorded in 5/9 tumours and 1/9 developed intracranial extension. No patients died during the follow-up period.

Patient 1 presented with 20/16 vision in the affected eye and was managed using observation alone. Follow-up after 89 months demonstrated no tumour growth and vision was 20/20. Patient 3 presented with 20/20 vision and was also treated conservatively.14 After 16 years, vision was 20/30 and the tumour had undergone cystic degeneration and decreased in size. This is the first reported case of radiological regression in PPONSM.

Patient 2 had bilateral PPONSM on presentation with vision of 20/16 in the right eye and light perception in the left eye. Both tumours involved the entire length of the intraorbital nerve. She was untreated for 11 years before her vision started to deteriorate. She developed no light perception in the left eye, as well as tunnel vision and reduced visual acuity of 20/80 in the right eye. Repeat imaging demonstrated volume growth of bilateral PPONSMs without intracranial extension. The patient was
treated with radiotherapy to preserve remaining vision in the right eye. She received 54 Gy of irradiation in 30 fractions over 6 weeks to the right ONSM. After the course of radiotherapy she experienced nausea, otalgia and local skin erythema. She was followed up for 8 years after radiotherapy. There was no further radiological growth of the right or left PPONSM and vision remained 20/80.

Patient 4 presented with light perception vision in the affected eye. She was observed for a period of 4 years when she developed intracranial tumour extension. Vision deteriorated to no light perception. MRI showed that the tumour had extended into the cavernous sinus and surrounded the internal carotid artery. She received 55.8 Gy of irradiation in 31 fractions over 6 weeks. After the course of radiotherapy, she experienced headaches periorbital oedema, fatigue and alopecia. The patient was followed up for 14 years after radiotherapy. There was no further radiological growth of the PPONSM and internal carotid artery flow was preserved. Vision remained no light perception.

Patient 6 had NF2 with multiple intracranial and spinal meningiomas. This patient presented with 20/25 vision in the affected eye. The patient underwent surgery for the intracranial and spinal tumours but not the PPONSM. Ten years after presentation, this patient experienced severe bilateral visual loss due to papilloedema. This occurred due to elevated intracranial pressure from the mass effect of recurrent NF2-associated intracranial tumours.

DISCUSSION
The incidence of intracranial meningioma is 2 in 100 000 per year for all age groups.17 Meningiomas generally occur in the fifth to eighth decades.19 Patients younger than 20 years account for 4% of all ONMS.1 Walsh19 first studied PPONSM in detail in 1970. The largest reports since 1970 were published by Karp et al20 in 1974 (10 patients), Saeed et al21 in 2003 (six patients) and Lee et al22 in 2008 (14 patients). We have identified 70 previously documented cases of PPONSM in the English-language medical literature (table 2). Other cases have been documented within larger reports of adult ONSM.23-25 However, these reports have not disclosed the numbers of patients younger than 20 years. Also, the patient numbers reported in the following sections may vary since some of the information sought was not disclosed in the literature. The sex distribution of PPONSM is 44 (64%) female patients and 25 (36%) male patients.

Association with NF2
There is a well-recognised relationship between NF2 and meningioma.26 The incidence of NF2 in PPONSM is greater than in adult ONSM.2 When our series was combined with reported cases, 23 out of 78 patients (29%) had concomitant diagnoses of NF2 and PPONSM (table 3). In many of these cases, PPONSM was the first manifestation of NF2. The majority of patients with PPONSM and NF2 were treated: 11 patients were treated surgically, two patients were treated with radiotherapy and two patients were treated with surgery followed by radiotherapy. Three patients, including two from this series, were observed without any treatment.

Smece et al28 presented a case of a 7-year-old boy with bilateral PPONSM who was treated surgically but was experiencing radiological tumour growth. This patient was later treated with radiotherapy then followed up for 5 years and showed no further tumour growth. Kiratli et al47 reported a 12-year-old girl who presented with light perception vision in the right eye and was found to have right PPONSM as well as a left schwannoma. The patient also had two intraretinal astrocytic hamartomas, bilateral cerebellopontine angle tumours, multiple cerebello-cortical tumours and a right parietal parafalcine meningioma. The patient was treated with radiotherapy and followed up for 12 months and her vision remained stable. Vanikieti et al29 reported a 12-year-old boy who presented with count fingers vision and was found to have right PPONSM. This patient also had a meningioma along the left anterior temporal convexity, a falci meningioma and multiple schwannomas of bilateral cranial nerves V, VI, VII, VIII, IX and XI, and spinal nerve roots. The patient was treated with radiotherapy and followed for 6 months and his vision remained stable. There are no reports with follow-up longer than 12 months in patients with NF2 treated with radiotherapy for PPONSM. The long-term safety of radiotherapy has not been established. There remains a potential risk of radiotherapy induced new tumour formation and malignant transformation of existing tumours.30-32

Natural history
Walsh19 first suggested that ONMSs were more aggressive in children than adults. Early medical literature therefore supported surgical resection.19,33 Visual outcomes were invariably poor. Recurrence of PPONSM after surgical resection has been reported by Walsh,19 Karp et al20 and Mallucci et al34. Walsh19 also reported a case of a 11-year-old girl with NF2 who presented with left PPONSM and underwent surgery, but later developed papilloedema in the contralateral eye due to a new intracranial tumour.

Prior to this case series, there had only been one reported case of PPONSM managed using observation alone.9 Lee et al8 reported an 18-year-old woman with NF2 who presented with 2 mm proptosis in the left eye and 20/20 vision bilaterally. The tumour was observed for 13 years and the patient had no change in vision and no tumour growth on MRI.

There are two additional reports of patients who were observed for several years before treatment. Walsh19 described an 11-year-old boy who was untreated for 17 years and developed decreased vision in the contralateral eye due to chiasmal tumour extension. Wright35 reported a 10-year-old boy with NF2 who presented with a 5-year history of poor vision. The patient was observed for 8 years when vision reduced to light perception and he developed bilateral acoustic neuromas. Prior to this study, the mean period of observation, from onset of symptoms to commencement of treatment, was 3.5 years ± 4.2 years (range: 1 month to 17 years; median: 2.5 years).

In our series, two (2.5%) out of eight patients presented with normal vision and were not treated. These patients had minimal change in vision and did not experience tumour growth after long-term follow-up. Out of our six untreated patients, four (67%) were not treated for PPONSM and experienced deterioration of their vision after long-term follow-up. Therefore, including the case reported by Lee et al8, a total of three patients in the literature have been found to have indolent tumours.

There have been six documented deaths in patients with PPONSM. All deaths resulted from operative complications or secondary causes.19,20,31 None of the deaths were attributed directly to PONSM. Three of these six patients had NF2 and developed other intracranial tumours. These patients all died from surgical complications following removal of the intracranial tumours. This highlights the difficult management problem presented by PPONSM in NF2. Most patients with NF2 face substantial morbidity and reduced life expectancy due to the development of other CNS tumours. For example, patients with NF2...
Table 2  Clinical details of previously reported cases of PPONSM

<table>
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<tr>
<th>Author</th>
<th>No of patients</th>
<th>Age (years)</th>
<th>Males</th>
<th>Females</th>
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<th>No of females</th>
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<th>Disc atrophy</th>
<th>Field defect</th>
<th>Intracranial disease</th>
<th>Treatment</th>
<th>Intervals of follow-up</th>
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<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Surgery</td>
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NF2, neurofibromatosis type 2; PPONSM, primary paediatric optic nerve sheath meningioma; VA, visual acuity.
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<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Side</th>
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<th>Disc findings</th>
<th>Other tumours</th>
<th>Treatment</th>
<th>Follow-up</th>
<th>Mortality</th>
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<td>11</td>
<td>F</td>
<td>L</td>
<td>V, R, S</td>
<td>12/200</td>
<td>A</td>
<td>Sacral neura, left and right cerebellopontine angle neuromas, multiple intracranial dural meningiomas</td>
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<td>Died age 16</td>
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<td>R</td>
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<tr>
<td>Mallucci et al</td>
<td>F</td>
<td>P</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Multiple acoustic neuromas, CN V neuromas, pansagittal meningioma after 11 years</td>
<td>Total surgical resection (then recurrence) then radiotherapy</td>
<td>14 years</td>
<td></td>
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<tr>
<td>Bosch et al</td>
<td>14</td>
<td>F</td>
<td>R</td>
<td>V, S</td>
<td>20/70</td>
<td></td>
<td></td>
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<tr>
<td>Lee et al case 2</td>
<td>18</td>
<td>F</td>
<td>P</td>
<td></td>
<td>20/20</td>
<td></td>
<td>Multiple CNS meningiomas</td>
<td>None</td>
<td>9 years</td>
<td></td>
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<tr>
<td>Lee et al case 4</td>
<td>13</td>
<td>F</td>
<td>V, P</td>
<td></td>
<td>20/400</td>
<td>O</td>
<td></td>
<td>Subtotal resection then enucleation</td>
<td>13 years</td>
<td></td>
</tr>
<tr>
<td>Lee et al case 5</td>
<td>14</td>
<td>M</td>
<td>P</td>
<td></td>
<td>20/20</td>
<td>O</td>
<td></td>
<td>Subtotal resection</td>
<td>9 months</td>
<td></td>
</tr>
<tr>
<td>Lee et al case 10</td>
<td>17</td>
<td>F</td>
<td>V, P</td>
<td>PL</td>
<td></td>
<td>A</td>
<td></td>
<td>Subtotal resection</td>
<td>13 years</td>
<td></td>
</tr>
<tr>
<td>Kiati et al</td>
<td>12</td>
<td>F</td>
<td>R</td>
<td>V, P</td>
<td>PL</td>
<td>O</td>
<td>Left orbital schwannoma, 2 x intraretinal astrocytic hamartomas, bilateral cerebellopontine angle tumours, right parietal parafalcine meningioma</td>
<td>Radiotherapy, radiosurgery to schwannoma</td>
<td>18 years</td>
<td></td>
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<tr>
<td>Sme et al</td>
<td>7</td>
<td>M</td>
<td>B/L</td>
<td>V</td>
<td></td>
<td></td>
<td></td>
<td>Surgery then radiotherapy</td>
<td>5 years</td>
<td></td>
</tr>
<tr>
<td>Vanikieti et al case 1</td>
<td>12</td>
<td>M</td>
<td>R</td>
<td>V, R, E</td>
<td>CF</td>
<td>O, S</td>
<td>Multiple neurofibromas, left temporal meningioma, falci ne meningioma, multiple schwannomas of bilateral CN V, VI, VII, VIII, IX, XI and spinal nerve roots</td>
<td>Radiotherapy</td>
<td></td>
<td></td>
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</tbody>
</table>

A, disc atrophy; B/L, bilateral; CF, count fingers; CN, cranial nerve; CNS, central nervous system; E, limitation of eye movement; S, scotoma; F, female; L, left; M, male; O, disc swelling; ONSM, optic nerve sheath meningioma; p, proptosis; PL, light perception; R, right; S, optociliary shunt vessels; V, decreased vision; VA, visual acuity.
diagnosed prior to 1990 had only a 15-year life expectancy from diagnosis.3,15 Advancements in management and early diagnosis are improving outcomes, but many patients with multitudinous disease still die in the third and fourth decades of life.36

Intracranial extension of PPONSM is sometimes considered an indication for treatment. Twelve (27%) out of 44 patients among the cases reported in literature, including our eight patients, had intracranial disease at presentation. In patients treated using surgical resection, 5 (31%) out of 16 with no previous evidence of intracranial disease developed intracranial extension after treatment. In comparison, intracranial disease affects approximately 20% of adult cases.37 These data support the suggestion that ONSM may have a more aggressive growth pattern in children than adults.

Summary
We have presented eight new cases of PPONSM. Six patients in this series, including two patients with NF2, were observed for several years. To our knowledge, this is the largest PPONSM case series with long-term data on patients who were treated primarily by observation. Two out of eight patients presented with good vision and did not experience tumour growth after long-term follow-up. This highlights that the natural history of some PPONSMs may be favourable. We acknowledge that the rarity of this disease makes it difficult to draw general conclusions from this case series. We nonetheless suggest patients who present with good vision could be initially managed conservatively. A diagnosis of PPONSM should promote a search for evidence of NF2. Patients who experience disease progression and worsening of vision can be considered for radiation therapy. However, late complications of radiotherapy in children have not been studied in detail. Induction of secondary orbital and intracranial meningiomas after radiation therapy has been documented.38-42 The dose of radiation, the remaining lifespan of the patient and the risk of radiation-induced side effects, especially in patients with NF2, must all be considered when making management decisions.

Contributors
DS conceived the study. DN, KL, JC and GD initiated the design of the study. DN, GT, AF and EF helped with the implementation of the study. DN and GT did the data collection, analysis and interpretation. DN, GT, AF and EF performed the drafting of the manuscript. All authors contributed to revision of the manuscript and final approval of the version to be published.

Competing interests
GT is supported by ‘Filling the Gap’ of the University of Zurich, Zurich, Switzerland.

Patient consent
Detail has been removed from this case description/these case descriptions to ensure anonymity. The editors and reviewers have seen the detailed information available and are satisfied that the information back up the case the authors are making.

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REFERENCES


Natural history of primary paediatric optic nerve sheath meningioma: case series and review

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