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


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

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 PPONSM 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.

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KEYWORDS: optic Nerve; orbit; vision

PMID: 29146762 DOI: 10.1136/bjophthalmol-2017-310672