

CLINICOPATHOLOGICAL AND SURGICAL CHARACTERIZATION OF OPTIC NERVE-ASSOCIATED ATYPICAL MENINGIOMAS DEVELOPED IN THE ANTERIOR AND MIDDLE CRANIAL FOSSA

Claudia Florida Costea^{1, 2}, Laurentiu Andrei Blaj^{2, 3}, Dana Mihaela Turliuc^{2, 3*}, Vlad Liviu Hartie⁴, Amelian Madalin Bobu⁵, Andrei Ionut Cucu^{2, 6}

¹Department of Ophthalmology, Faculty of Medicine, Grigore T. Popa University of Medicine and Pharmacy, 700115 Iași, Romania; claudia.costea@umfiasi.ro (C.F.C.)

²Prof. Dr. Nicolae Oblu Emergency Clinical Hospital, 700309 Iași, Romania

³Department of Neurosurgery, Faculty of Medicine, Grigore T. Popa University of Medicine and Pharmacy, 700115 Iași, Romania; laurentiu-andrei.blaj@d.umfiasi.ro (L.A.B.), turliuc_dana@yahoo.com (D.M.T.)

⁴Department of Anesthesia and Intensive Care, Grigore T. Popa University of Medicine and Pharmacy Iași, 700115 Iași, Romania; liviu.vlad.hartie@umfiasi.ro (V.L.H.)

⁵Department of Cardiology, "Sf. Spiridon" University Hospital, 700111 Iasi, Romania; amelian.bobu@gmail.com (A.B)

⁶Department of Medical-Surgical and Complementary Sciences, Faculty of Medicine and Biological Sciences, University Stefan cel Mare of Suceava, 720229 Suceava, Romania; andrei.cucu@usm.ro (A.I.C.)

*Correspondence author: Dana Mihaela Turliuc (D.M.T.) turliuc_dana@yahoo.com

Abstract

Meningiomas developing in the anterior and middle fossa can compress the optic nerve (ON) and cause secondary visual disturbances. The aim of this study was to evaluate the clinical, pathological, and surgical characteristics of atypical meningiomas (WHO grade II) compressing the ON and located in the anterior and middle fossa of the skull base. *Materials and Methods:* We retrospectively studied the data of 5 patients with atypical meningiomas compressing the ON, diagnosed at the Prof. Dr. Nicolae Oblu Emergency Clinical Hospital in Iași, from 2010 to 2019. *Results:* Out of 5 patients, 4 presented with intracranial hypertension syndrome at admission, and 2 with motor deficits. Most patients (4 out of 5) had reduced visual acuity (VA) at admission, and 3 of them had partial or total ON atrophy. The preferred surgical approach was the pterional approach for sellar and parasellar meningiomas (2 cases), and the frontotemporal approach for large meningiomas with extension (3 cases). *Conclusion:* Although most patients had reduced VA at the time of admission, they presented to the hospital with other neurological symptoms. This suggests that the slow growth of intracranial meningiomas and the gradual compression of the ON may delay early diagnosis and, more importantly, effective treatment in these patients. We recommend regular evaluation of VA, as early detection of a slow-growing intracranial tumor, such as a meningioma, can help prevent vision loss.

Introduction

Meningiomas located in the anterior [1] and middle fossa [2-5] can affect vision by compressing the optic nerve (ON) in any of its segments - whether intraorbital, canalicular, or intracranial [6].

Intracranial meningiomas, particularly those located in the olfactory groove, planum sphenoidale, tuberculum sellae, diaphragma sellae, giant pterional or cavernous sinus regions, can compress the ON between the optic foramen and the optic chiasm [1-4, 6-8]. A prefixed chiasm may be a predisposing factor, whereas a postfix chiasm may delay the diagnosis of ON compression [9]. Another factor influencing

the early onset of visual symptoms is the environment surrounding ON. Thus, ON meningiomas located in the optic canal are more likely to cause visual impairment [10, 11].

Another factor influencing the onset of visual symptoms is the histopathology of the meningioma, particularly the tumor-nervous tissue relationship [6]. Some meningiomas displace the ON while preserving the arachnoid plane, whereas others invade the ON or grow entirely within the ON sheath in the absence of an arachnoid plane [12-14]. Regarding the histopathology of meningiomas, the benign to non-benign ratio is 5.3:1 [15]. In a 14-

year longitudinal study that mapped the spatial distribution of histological subtypes in 272 patients with intracranial meningiomas (WHO grades I, II, and III), Shrestha *et al.* observed that both benign and non-benign meningiomas were most common on the convexity, followed by parafalcine and cerebellopontine angles. When analyzing histological subtypes, Shrestha *et al.* found that grade II and III meningiomas were most commonly found in supratentorial locations [15]. In addition, Hale *et al.* observed that in 128 patients with intracranial meningiomas, tumors along the falx and convexity were more frequently atypical than those at the skull base, and of the 24 skull base meningiomas, only 9 (34.6%) were WHO grade II [16].

Over the past 30 years, the management of ON-associated meningiomas has evolved considerably [10, 13, 17-20]. Current treatment strategies for ON-associated meningiomas include observation, radiotherapy, and surgical resection [6]. However, the common goal of all three strategies, regardless of the location of compression along the ON, is to stabilize or improve vision in the affected eye. Furthermore, in patients with total blindness in the ipsilateral eye, preserving vision in the contralateral eye becomes the primary goal of the treatment [5]. In cases of large intracranial meningiomas, surgical treatment is recommended, as these cases threaten not only the vision but also the patient's life [1, 2]. In such cases, the optic canal can be unroofed during surgery if there are clear signs of invasion, especially since meningiomas are known to cause bony changes [21], and bony invasion is associated with a worse prognosis, particularly in atypical meningiomas [22].

However, the efficacy of optic canal unroofing in these cases remains controversial. The procedure carries a risk of worsening vision [23, 24], especially in recurrent meningiomas, where adhesions between the meningioma and the optic apparatus may lead to potential

postoperative visual impairment [25]. Currently, radiotherapy for ON sheath meningiomas is considered less risky for vision preservation compared to surgery [18, 24, 26-32].

The aim of this study was to analyze the clinical, pathological and surgical characteristics of five patients with ON-associated atypical meningiomas located in the anterior and middle fossa of the skull base.

Material and methods

Our retrospective study included 5 patients with ON-associated atypical meningiomas located in the anterior and middle fossa of the skull base. These patients were surgically treated and diagnosed at the *Prof. Dr. Nicolae Oblu* Emergency Clinical Hospital in Iași, Romania, and were followed up between January 1, 2010, and December 31, 2019. Statistical data analysis was performed using SPSS 24.0 software (SPSS, Inc., Chicago, IL, USA) for Windows.

Results

Our study included 5 patients with intracranial atypical meningiomas located in the anterior and middle fossa of the skull base, compressing the ON. Regarding patient gender, one was female and four were male. The average age was 56.9 years (range 40–67 years). Clinically, 4 out of the 5 patients presented with intracranial hypertension syndrome at the time of admission, and 2 of them had motor deficits due to the large size of the tumor. Regarding visual disturbances, 4 out of the 5 patients had reduced visual acuity (VA), and 3 of them had partial or total atrophy of the ON on the side of the tumor.

Regarding the surgical technique, due to the anatomical location of the meningiomas, the preferred surgical approach was the pterional approach (2 cases) or the frontotemporal approach for larger meningiomas (3 cases). In our study, none of the 5 patients died postoperatively,

and the neurological outcomes were favorable, with the resolution of preoperative motor deficits. No additional visual disturbances were observed

postoperatively. The main clinical, pathological, and surgical characteristics of the patients in the study are presented in the table below (Table 1).

Table 1. Clinical, pathological, and surgical characteristics of patients in our study (n=5)

Case	Gender/ Age (years)	Tumor location	Signs and symptoms at admission	Intracranial hypertension syndrome	Motor deficit	Surgical approach	Visual acuity	Optic nerve atrophy
1	F/40	Sellar tuberculum	Left-sided anosmia	+	-	Pterional	Normal in both eyes (5/5)	No presence
2	M/64	Sellar diaphragm	Decreased LE visual acuity	-	-	Pterional	RE- normal (5/5) LE low (<5/100)	Total
3	M/53	Cavernous sinus (L)	Seizures	+	-	Frontotemporal	RE- normal (5/5) LE - low	Partial
4	M/60	Cavernous sinus (L)	Right hemiparesis	+	+	Frontotemporal	RE- normal (5/5) LE - low (9/10 BCVA)	No presence
5	M/67	Pterional (L)	Confusion Aphasia	+	+	Frontotemporal	OU low - (2/10 BCVA)	Partial

*F- female, M-male, L-left, LE-left eye, RE-right eye; BCVA – best corrected visual acuity; OU – both eyes

Case 1

A 40-year-old female patient was admitted to our service for persistent headaches and left-sided anosmia. Upon admission, her VA was normal, and the ON examination did not reveal any pathological changes. The head MRI scan revealed a sellar tuberculum meningioma (Figure 1), and surgery was performed via a left pterional approach, with gross total resection of the tumor. The postoperative course was favorable.

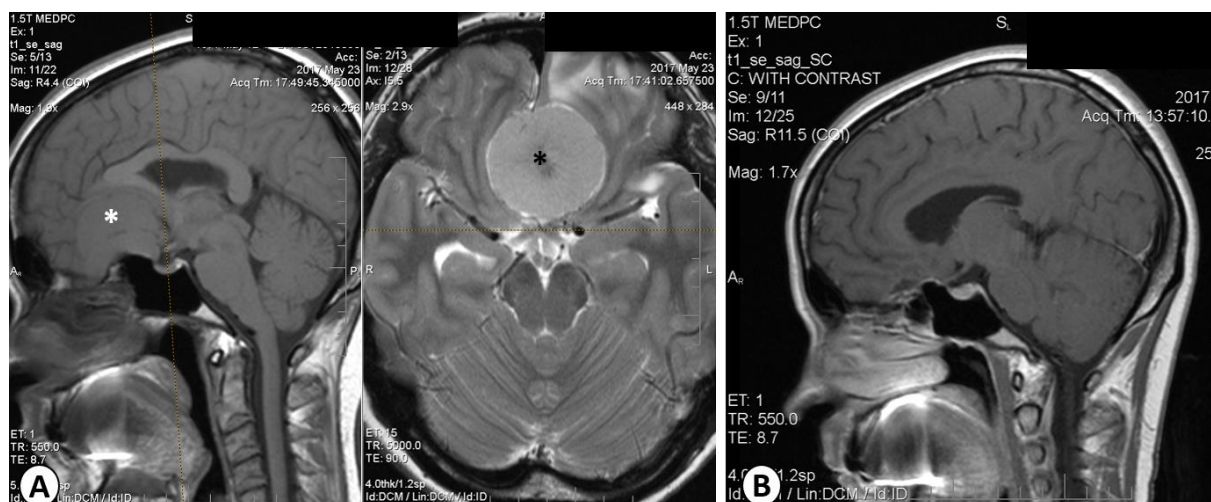


Figure 1 (A). Sagittal T1W and axial T2W MRI showing tuberculum sellae meningioma (black and white asterisk); **(B)** Postoperative axial contrast-enhanced T1W MRI showing the gross total resection. *MRI- magnetic resonance imaging, T1W - T1-weighted.*

Case 2

A 64-year-old male patient was admitted due to a dramatic decrease in VA in the left eye and progressive constriction of the visual field in the right eye, without any other complaints. The contrast-enhanced head MRI revealed a sellar diaphragmatic meningioma (Figure 2).

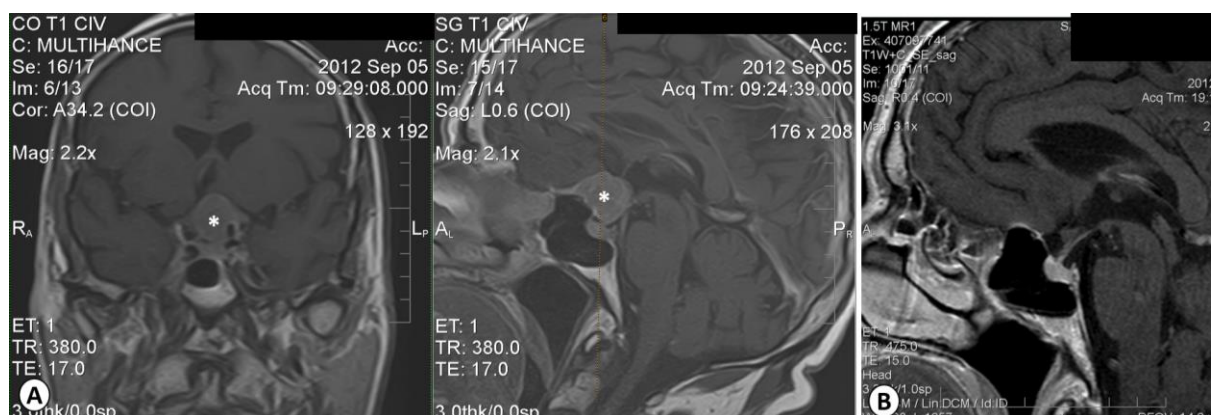


Figure 2 (A). Coronal and sagittal contrast-enhanced T1W MRI showing meningioma of diaphragm sellae (white asterisk); **(B)** Postoperative sagittal contrast-enhanced T1W MRI showing complete resection. *MRI – magnetic resonance imaging; T1W – T1-weighted.*

Ophthalmic examination revealed decreased VA in the left eye (<5/100) along with atrophy of the left ON. The head CT scan also identified compression of both optic nerves, especially the left ON (Figure 3).



Figure 3. Coronal and axial contrast-enhanced CT showing a diaphragm sellae meningioma (black asterisk), compressing the bilateral ONs, especially the left ON. *CT- computer tomography, ON – optic nerve, ONs – optic nerves.*

Surgery was performed, with resection of the tumor and decompression of the optic chiasm and pituitary stalk through a left pterional approach. The postoperative course was marked by a syndrome of inappropriate ADH secretion (SIADH), which subsequently resolved.

Case 3

A 53-year-old male patient was admitted for the sudden onset of seizures. Ophthalmic examination on admission revealed decreased VA in the left eye, along with partial atrophy of the left ON. Head MRI showed a giant meningioma of the left cavernous sinus with extension

into the left middle cranial fossa (Figure 4). The tumor was subtotal resected through a left frontotemporal approach. On the fifth postoperative day, the patient developed a stroke, which subsequently resolved.

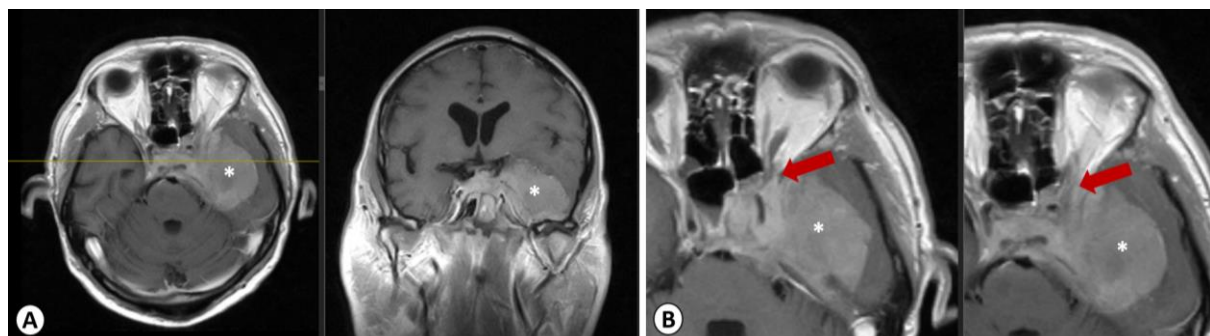


Figure 4 (A). Axial and coronal contrast-enhanced T1W MRI showing a giant left cavernous sinus meningioma, with extension into the middle fossa (white asterisk); **(B)** The same tumor (white asterisk), with compression of the left optic nerve (red arrow). *MRI –magnetic resonance imaging, T1W-T1 weighted.*

Case 4

A 60-year-old male patient was admitted to our neurosurgical service with a syndrome of intracranial hypertension and right-sided hemiparesis. The patient had no significant pathological history, being known only for left anterior fascicular block. Ophthalmic examination revealed decreased VA (9/10 BCVA) in the left eye, but with a normal appearance of the ON. Head MRI showed a left cavernous sinus meningioma with invasion of the inner third of the sphenoid wing and the temporo-polar basal area (Figure 5).



Figure 5 (A). Axial and coronal contrast-enhanced T1W MRI showing a left cavernous sinus meningioma. **(B)** Axial T2W MRI showing compression and displacement of the left optic nerve by the tumor. *MRI –magnetic resonance imaging, T1W – T1 weighted.*

A gross total resection of the tumor was performed (Figure 6) through a left frontotemporal approach, with a favorable postoperative course.

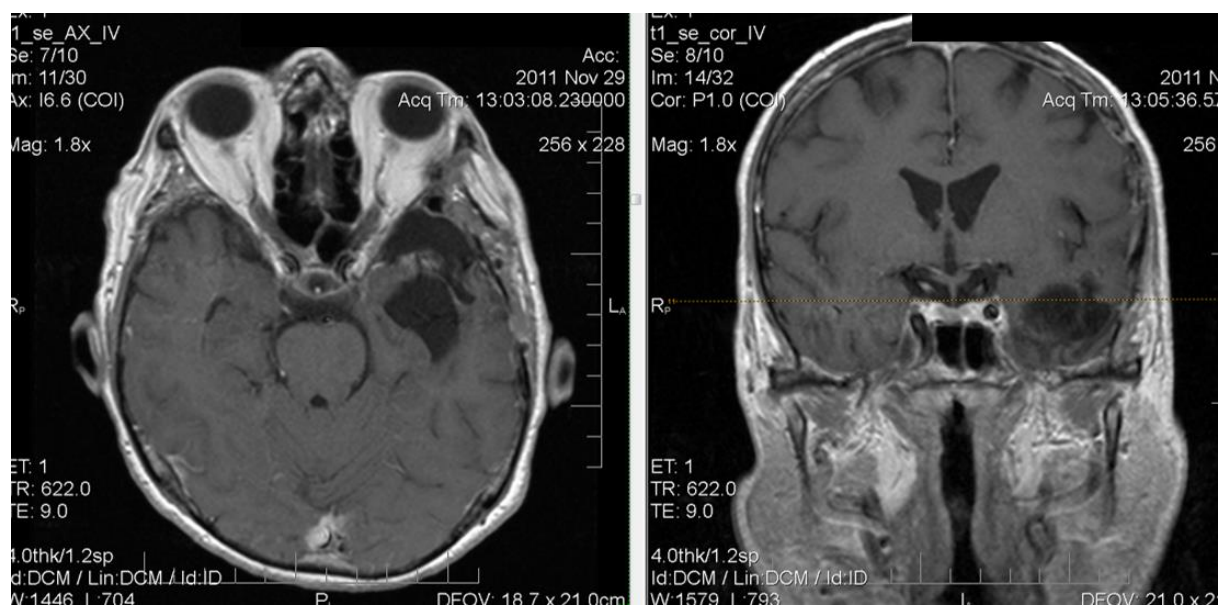


Figure 6. Postoperative axial and coronal axial contrast-enhanced T1W MRI showing the gross total resection of the tumor. *MRI –magnetic resonance imaging, T1W – T1 weighted.*

Case 5

A 67-year-old male patient was admitted for confusion and aphasia and right-sided hemiparesis with a sudden onset over the past few weeks. Ophthalmic revealed partial atrophy of the left ON. Head MRI showed a giant left pterional meningioma (Figure 7), which was surgically resected through a left frontotemporal approach with gross total resection. The postoperative course was favorable, with resolution of the confusion, aphasia and motor deficit.

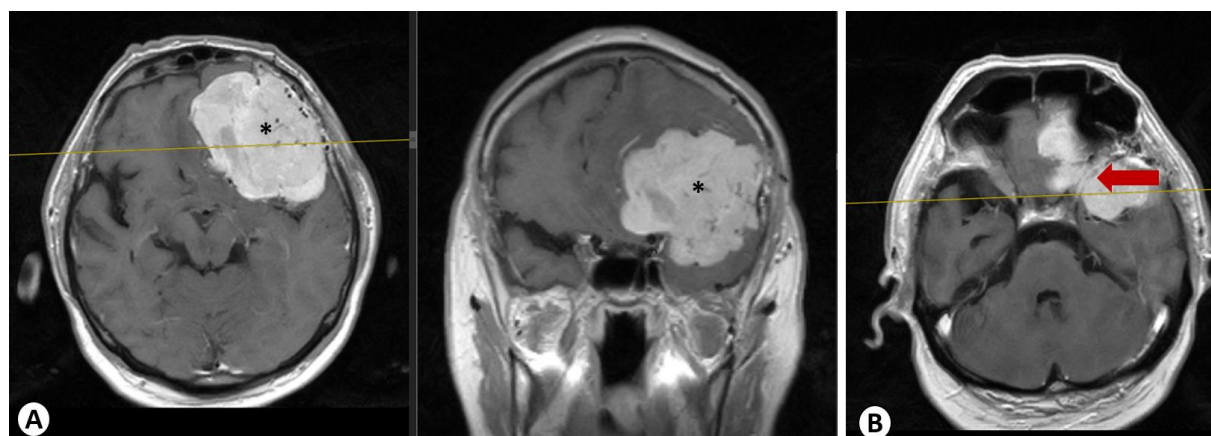


Figure 7. (A) Axial and coronal contrast-enhanced T1W MRI showing a left pterional meningioma (black asterisk); (B) The same tumor (black asterisk) compressing the entry of the left ON in the skull (red arrow). *MRI –magnetic resonance imaging, T1W – T1 weighted.*

Discussions

With regard to ON compression, the authors report three groups according to the location of the ON compression: (i) isolated compression of the ON within the orbit, (ii) compression of the ON within the optic canal, and (iii) isolated compression of the ON in the intracranial space [6].

In a systematic review that included meningiomas compressing the ON, Hénau *et al.* found that out of 317 patients, 154 had ON compression in the intracranial space (6). Of these, 90% ($n=138$) presented with preoperative visual disturbances. Similarly, in our study, of the 5 patients with atypical meningiomas compressing the ON, 4

presented with decreased VA on admission, and 3 showed partial or total ON atrophy (2 patients with partial atrophy and one with total ON atrophy). The mean age was 56.8 years (range 40-67 years), similar to the results obtained by Hénau *et al.*, where the mean age was 53 years [6].

In our patient cohort, the preferred surgical approach was the pterional approach, with its frontal and temporal extensions, which are used for giant meningiomas. To date, the literature classically reports the pterional approach as conventional and preferred for sellar and suprasellar tumors [33], and it remains a preferred approach even when compared to more invasive skull base approaches [11]. Advocates of the pterional approach also support it because it is a fast, non-aggressive, and well-known approach among neurosurgeons [6]. Regarding extradural anterior clinoidectomy [34], or the extradural opening of the optic canal, although this allows easier mobilization of the ON, it is not without surgical morbidity, particularly due to the increased risk of cerebrospinal fluid fistulas, owing to the potential pneumatization of the anterior clinoid process.

Postoperatively, most patients experienced no visual improvement due to the irreversible changes in ON atrophy, which was present in 3 out of the 5 patients.

In the literature, based on the 2016 WHO classification, atypical meningiomas (WHO grade II) account for 16.9% of cases [35, 36]. In a study that examined the spatial distribution of histological subtypes of meningiomas over a 14-year period in 272 patients, it was found that out of 36 patients with atypical meningiomas, one (0.4%) was located in the suprasellar region, one (0.4%) in the clinoid region, and 8 (2.9%) in the sphenoid wings. On the other hand, the majority of atypical meningiomas were located in the convexities ($n=11$, 4%) and 9

in the parasagittal/parafalcine regions [15]. This further emphasizes the rarity of atypical meningiomas located in the sellar and parasellar regions. Regarding cavernous sinus meningiomas, in a study where He *et al.* retrospectively followed 54 patients with cavernous sinus meningiomas, found that 20% of them ($n=10$) had a diagnosis of WHO grade II meningiomas [37].

Regarding the residual tumor rate, studies have reported a rate of 25% for tumors invading the optic canal and 39% for intraorbitally located tumors. However, only one in two patients received postoperative radiotherapy [6]. These high rates of residual tumors may be explained by the preoperative failure to recognize tumor extension into the optic canal, which may go undetected during surgery. Secondly, the increasing role of radiosurgery in skull base surgery [18, 27, 30, 32] has led to a paradigm shift in treatment strategies for benign skull base tumors. Consequently, decision-making in skull base surgery is now focused more on patient safety than on the complete tumor removal [38, 39].

Conclusions

In our series of atypical meningiomas with ON compression, we observed that although most patients had reduced VA on admission, they presented with other neurological symptoms. Of the 5 patients, only one presented with visual disturbances because they were extremely severe due to total ON atrophy. This finding suggests that the slow growth of intracranial meningiomas and gradual compression of the ON can delay early diagnosis and effective treatment for these patients. Periodic VA examinations could be useful for early diagnosis and prevention of vision loss in patients with tumors compressing the optic nerves.

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