Spontaneous recovery of Medial Prefrontal Syndrome following Giant Olfactory Groove Meningioma resection: A case report

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ABSTRACT

Introduction: Anterior skull base meningiomas are frequently associated with changes in personality and behavior. Olfactory groove meningiomas take their origin from the cribiform plate and frontoethmoidal suture in the floor of the anterior cranial base. The cardinal clinical features usually observed are mental changes, visual deterioration, and headaches as a result of olfactory or optic nerve and frontal lobe compression. However, several authors have reported minimal changes in cognition or behavior following surgical resection, and many patients do not return to their premorbid level of function.

Case presentation: A 42-year-old male with four months history of changes in behavior, apathy, and lack of motivation. Over the course of two months, his vision progressively worsened in both eyes, and he developed a diffuse, mild headache. On examination, he was fully alert and cooperative. He only had light perception and could see faint silhouettes of people. All other neurological examinations were normal. Head Computed Tomography (CT) demonstrated a large, extra-axial mass consistent with an anterior skull base mass extending to the sellar and suprasellar region, which suggests an olfactory groove meningioma. The patient underwent a bifrontal approach craniotomy tumor resection and subsequently recovered from medial prefrontal syndrome following tumor resection. Surgical tissue pathology confirmed the diagnosis as an olfactory groove meningioma.

Discussion: The anatomical location of the olfactory groove meningioma may cause prolonged psychiatric symptoms before the onset of more overt neurological deficits. Personality changes that occur may be subtle, so patients may avoid seeking treatment for their headaches and delay ophthalmic care until their complaints become severe. Anterior skull base meningiomas often involve the ventromedial prefrontal cortex (vmPFC) that is associated with elevated levels of apathy.

Conclusion: Olfactory groove meningiomas are treatable. Reversal of cognitive impairments and changes in behavior can be expected after surgical resection.

Keywords: Complete recovery, giant olfactory groove meningioma, medial prefrontal syndrome, total removal.


INTRODUCTION

Meningiomas are the most commonly described intracranial tumors, accounting for 36% of all primary brain neoplasms in adults.¹ Anterior skull base meningiomas are frequently associated with changes in personality and behavior.² Olfactory groove meningiomas originate from the cribiform plate and frontoethmoidal suture in the floor of the anterior cranial base.³ The incidence of olfactory groove meningioma is approximately 4 –13% among all intracranial meningiomas.⁴ Olfactory groove meningiomas are slow-growing tumors, often bilateral rather than unilateral, but asymmetrical in growth. They usually remain clinically latent during the early phases of tumor growth, leading to large tumor sizes at the time of diagnosis. They may produce progressive compression of the frontal lobes and project backward towards the sella. If large enough, they can affect vision by compressing the optic nerve and chiasm.⁵

The cardinal clinical features usually observed are mental changes, visual deterioration, and headaches as a result of olfactory or optic nerve and frontal lobe compression.⁶ Most often, patients present with psychiatric symptoms, such as apathy, depression, or psychomotor retardation, whereas urinary incontinence and gait disturbances are late findings. Consequently, patients often receive psychiatric treatments, sometimes for prolonged periods, before neurologic deterioration prompts imaging of the brain; at the time of diagnosis, most tumors are very large (> 4 cm).⁷

Anterior skull base meningiomas often involve the ventromedial prefrontal cortex (vmPFC), which is involved in various higher cognitive functions (including value-based decision-making and moral judgments) and also in emotional regulation and personality.³ Interestingly, although anterior skull base meningiomas frequently involve the vmPFC, the cognitive and behavioral manifestations of
CASE REPORT

Anterior skull base meningiomas remain poorly understood. Several authors have reported minimal changes in cognition following surgical resection of meningiomas. However, despite no significant measurable difference in cognitive or behavioral functioning after surgery, many patients do not return to their premorbid level of function.

Numerous surgical approaches have been recommended to tackle olfactory groove meningiomas, with the fronto-lateral, pterional and subfrontal (unifrontal or bifrontal) and their variations being the most common open transcranial procedures. However, describing the ideal surgical approach is still controversial.

We present a case of a giant olfactory groove meningioma encompassing the entire frontal lobe and compressing the optic nerves and chiasm, causing mental changes. The subsequent recovery from medial prefrontal syndrome following tumor resection is also presented.

CASE PRESENTATION

A 42-year-old male with four months history of changes in behavior, apathy, and lack of motivation was referred to a neurosurgeon. Over the course of two months, his vision progressively worsened in both eyes, and he developed a diffuse, mild headache. The following symptoms were absent: seizures, vomiting, diplopia, hallucinations, decreased sense of smell and taste, agitation, motoric and sensory loss. On examination, he was fully alert and cooperative. He only had light perception and could see faint silhouettes of people. All other neurological examinations were normal.

An emergent non-contrast computerized tomography (CT) image of the head (Figure 1) revealed a large extra-axial mass, approximately 6.6 × 6.3 cm, hyperdense, and partially calcified extending from the planum sphenoidale and cribriform plate to the upper frontal convexity into the sellar/suprasellar region, with signs of peritumoral edema, optic chiasm, and bilateral intracranial optic nerves compression.

Head CT with contrast images (Figure 2) further delineated non-contrast CT findings and highlighted a large avidly enhancing midline extra-axial mass within the frontal region, measuring approximately 6.6 × 6.1 cm in axial dimension, consistent with a meningioma. The mass extended inferiorly to the cribriform plate and into the sellar/suprasellar, compressing the optic chiasm and bilateral intracranial optic nerves.

The patient underwent a bifrontal approach craniotomy with tumor resection and pericranial graft. The patient was discharged from the hospital on the fourth day after surgery. Surgical tissue pathology confirmed the diagnosis as an olfactory groove meningioma (Meningothelial meningioma; WHO grade 1) (Figure 3).

At one-week follow-up after surgery, the patient recovered with cognitive/behavioral improvement, persistent poor vision, and normal subjective sense of smell. He became excited and passionate about doing his daily activities. The patient did not experience any abnormal movements or paresis. Three weeks postoperative head CT with contrast images (Figure 4) showed a post-operative cavity with extensive fluid and extra-axial pneumocephalus in the surgical bed, with no evidence of residual tumor.

DISCUSSION

Olfactory groove meningiomas most commonly present with symptoms of headaches, anosmia, or personality changes. The anatomical location of the olfactory groove meningioma may cause prolonged psychiatric symptoms before the onset of more overt neurological deficits. Due to these subtle symptoms before clinical presentation, olfactory groove meningiomas can grow insidiously large and present as one of the largest intracranial tumors. Tumor size has been categorized by neuroimaging evaluation. The new classification system was developed using the tumor size and anterior fossa extension as parameters (Figure 5). Type I (small) tumors are localized to 1 segment and are sized < 2 cm. Type II (moderate) tumors are localized to 2 segments and are sized 2 – 3.9 cm. Type III (large) tumors are localized to 3 segments and are 4 – 5.9 cm in size. Type IV (giant) tumors are localized to 4 segments and are > 6 cm in size.

Our current case, which was a giant olfactory groove meningioma measuring 6.6 cm in axial diameter, emphasizes the huge tumor burden that can be present before neurological symptoms.

Figure 1 Axial CT Head non-contrast images demonstrating a 6.3 cm transverse of 6.6 cm AP extra-axial mass. Edematous changes are seen within the frontal lobes. The mass occupies the anterior cranial fossa floor. The mass appears to extend over the right-sided sphenoid ridge, might extend over the left sphenoid ridge, and is likely to extend into the sella/suprasellar.
CASE REPORT

According to several large retrospective case reviews of olfactory groove meningioma, the most common presenting symptom is a headache, associated with large and giant tumors, followed by mental status alteration and seizures. Visual symptoms usually occur after an olfactory groove meningioma has reached a considerable size, but this can depend on their exact origin. Olfactory groove meningiomas extend posteriorly; this extension can cause the compression of one or both of the optic nerves or the chiasm and may occur earlier in smaller tumors arising from the posterior half of the cribriform plate. Compression injury to the optic nerves may induce optic nerve head edema, which may eventually progress to optic atrophy, and can also present as a Foster–Kennedy syndrome. The mechanism of injury in an olfactory groove meningioma is due to the large size of the tumor, which places downward pressure on the nerve and chiasm from above, unlike a suprasellar meningioma, which elevates the chiasm, thereby putting the crossed fibers on stretch and causing bitemporal hemianopsia with optic atrophy.

In the current report, our patient reported a four months history of changes in behavior, apathy, and lack of motivation. Over the past two months, he experienced a gradual worsening of headaches along with visual complaints, which was likely due to the giant nature of his olfactory groove meningioma. As personality changes can be subtle, patients may avoid seeking treatment for their headaches and delay ophthalmic care until their complaints become severe. Apathy must be distinguished from major depressive disorder and chronic fatigue syndrome. When patients presenting with apathy are asked about their mood, they state that they are not depressed, but instead they are experiencing chronic fatigue and lack of motivation. This may be associated with a functional disconnection between the frontal lobe and paralimbic areas, or damage in these areas. The diagnostic criteria for apathy include lack of motivation, diminished goal-directed behavior (lack of effort or dependency on others to structure their activity), diminished goal-directed cognition (lack of interest in learning new things or in new experiences, or lack of concern about one’s personal problems), or diminished emotions (unchanging effect, or lack of emotional responsivity to positive or negative events).

Anterior skull base meningiomas often involve the vmPFC, which is implicated in various higher cognitive functions (including value-based decision-making and moral judgments) and also in emotional regulation and personality. The vmPFC damage was associated with elevated levels of apathy. The prefrontal behavioral symptoms...
could also be divided into two syndromes, namely the cingulate/prefrontal medial syndrome and the orbitofrontal syndrome. The cingulate/prefrontal medial syndrome is characterized by apathy, withdrawal, decreased communication, and depression, while the symptoms of orbitofrontal syndrome include impulsivity, disinhibition, personality disorders, aggressiveness, and other similar manifestations. Medial frontal lobe damage causes apathy or abulia (a severe form of apathy). Acute bilateral lesions in the medial frontal area can cause akinetic mutism, in which the individual is awake and has self-awareness, but does not initiate behaviors.**

The ventromedial areas of the prefrontal cortex are involved in the expression and control of emotional and instinctual behaviors. Furthermore, the cingulate gyrus (medial frontal region) is also closely related to the emotional/motivational dysexecutive syndrome. Damage to the anterior cingulate gyrus causes apathy or abulia.

Olfactory groove meningioma can be surgically removed through a number of surgical techniques, namely the bifrontal approach, the unilateral frontal approach, the unilateral pterional-transylvian approach, the pterional approach, the frontotemporal approach, the lateral supraorbital approach, and minimally invasive endoscopic techniques. All approaches have both their advantages and disadvantages, but the bifrontal approach allows for good exposure to the giant olfactory groove meningioma and the ethmoidal area. Our patient underwent the bifrontal approach craniotomy with complete resection of his giant olfactory groove meningioma. Due to the extensive size of his tumor, this approach was deemed to be more successful in removing the entire tumor and decreasing the likelihood of recurrence than other approaches.

Several authors have reported minimal changes in cognition following the surgical resection of meningiomas. However, despite no significant measurable difference in cognitive or behavioral functioning after surgery, many patients do not return to their premorbid level of function. Nevertheless, our patient recovered with cognitive/behavioral improvement at one-week follow-up after surgery. He became excited and passionate about doing his daily activities. The importance of this case report is that it illustrates the changes in behavior, apathy, and lack of motivation that can occur as a result of insidious tumors, such as olfactory groove meningiomas. These slow-growing, giant tumors can encompass the entire anterior skull base, causing minimal symptoms, such as personality changes, mild headaches, visual loss, and anosmia. However, the personality changes experienced by patients may be subtle, so patients may avoid seeking treatment for their headaches and delay ophthalmic care until their complaints become severe. Olfactory groove meningiomas can be successfully treated surgically, with cognitive/behavioral improvement after surgery.

**CONCLUSION**

Olfactory groove meningiomas are treatable. Symptoms and signs of cognitive impairments and behavioral changes suggest a frontal lobe lesion; this together with headaches, visual symptoms and anosmia should raise the suspicion of an olfactory
groove meningioma. Following surgical resection, reversal of the changes in thought processes can be expected.

CONFLICT OF INTEREST
The authors declare no conflict of interest regarding this manuscript.

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REFERENCES