

# Case report of a rare case of primary sinonasal meningioma

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The aim of this study was to report a case of sinonasal meningioma in Sudan. Meningiomas account for nearly 20% of all intracranial neoplasms and are the second most common tumor of the central nervous system. Primary meningiomas of the nose and paranasal sinuses are extremely rare. The clinical and radiological features of these tumors are nonspecific, and consequently an accurate diagnosis requires histologic evaluation. A 62-year-old woman from North Sudan presented to our hospital in May of 2016 complaining of right nasal obstruction associated with bloody nasal secretion and proptosis. Endoscopic endonasal examination revealed a soft mass in the right nasal cavity between the septum and the middle and superior turbinates posteriorly. MRI revealed the presence of a lobulated mass with soft-part density in the right nasal cavity (middle meatus) and the right ethmoid sinus invading the orbital contents and extending intracranially. The biopsy of the lesion revealed nodular neoplasm composed of regular cells arranged in whorled pattern. One psammoma body was noted and there was no evidence of pleomorphism or necrosis. The features are consistent with sinonasal meningioma. The patient was referred to the oncological department because it was an unresectable malignant meningioma and surgery was not feasible and would create more complications. Meningiomas involving the nasal cavity and paranasal sinuses are rare. The mortality is low, and its lethality is due to the complications of the surgery and injury of vital structures. Complete surgical extirpation of sinonasal tract meningiomas has an overall good prognosis. Hence, endoscopic nasal approach is an excellent surgical option in these cases, due to its low morbidity and satisfactory lesion resection.

### Keywords:

meningiomas, nasal cavity, paranasal sinuses

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### Introduction

Meningioma is an ordinary intracranial neoplasm, with a histomorphological development standard and, in general, easily recognized. Therefore, primary extracranial meningiomas of the nasal cavity, the paranasal sinus, and the nasopharynx are rare. The literature and its review are usually limited as regards isolated case reports [1].

Meningiomas account for nearly 20% of all intracranial neoplasms; primary meningiomas of the paranasal sinuses are extremely rare. Only 30 cases have been previously reported [2].

Meningiomas arise from meningotheial arachnoid cells, which derive from neuroectodermal tumors. Therefore, they are usually classified as mesoderm tumors [2].

Seventy-six of tumors have progesterone receptors, 96% have somatostatin receptors, 89% have epidermal growth factor receptors, and 19% have estrogen receptors. Hence, the use of tamoxifen and RU-486 (antiprogesterone) are under study. Characteristic and most frequent chromosomal aberration in meningiomas is monosomy 22, which, however, has been shown not to be relevant to the prognosis. Radical surgical resection remains the

primary mode of treatment and is correlated with a good prognosis. Efficacy of adjunctive radiotherapy after surgery is not established. Radiotherapy is used in unresectable malignant meningioma or recurrent meningioma for which surgery is not feasible. Newer techniques in treatment include proton irradiation and stereotactic radiosurgery with gamma knife [3,4].

### Case report

A 62-year-old Sudanese woman presented to Africa ENT specialized hospital in Sudan in May 2016 complaining of right nasal obstruction associated with mucoid bloody secretion and proptosis. Four weeks later the patient developed blindness on the right side and headache. Endoscopic endonasal examination revealed a soft mass in the right nasal cavity between the septum and the middle and superior turbinates posteriorly. MRI revealed the presence of a lobulated mass with soft-part density in the right nasal cavity (middle meatus) and the right ethmoid sinus and invading the orbital contents engulfing the

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optic nerve and extended intracranially. The biopsy of the lesion was obtained under general anesthesia by means of endoscopic endonasal surgery and it revealed nodular neoplasm composed of regular cells arranged in whorled pattern. One psammoma body was noted and there was no evidence of pleomorphism or necrosis; the features are consistent with sinonasal meningioma (Figs. 1–4).

**Discussion**

Meningioma is a tumor of the central nervous system, but it has rarely been occurring as a primary extracranial tumor of the paranasal sinuses [2].

Meningiomas affecting the paranasal and the nasal cavity are rare. They occur as a result of an extension of a primary intracranial mass, affecting only around 3% of

the cases, and hence it not a common event. Such tumors can also be reported with some evidence of intracranial tumor. Their origin is uncertain, including the origin of arachnoid cells around nerves or heterotopic cells when displaced in the moment of the structure closing of the embryo’s midline. The origin of Schwann cells or mesenchymal pluripotent ones is also suggested [5].

A variety of signs and symptoms are reported as regards clinical terms, including nasal obstruction and secretion, nasal polyps, maxillary pain, periorbital edema, exophthalmia, proptosis, ptosis, sight reduction, hearing loss, and headache [5].

The classification system of Hoyer encompasses the major etiologies proposed in the development of extracranial meningiomas [6]:

**Figure 1**



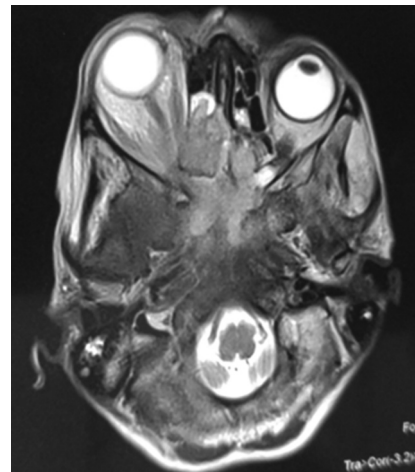
Rt eye proptosis and displacement.

**Figure 3**



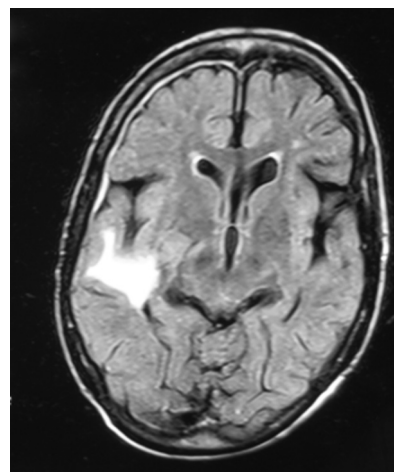
MRI coronal, showed a mass in the Rt sphenoid sinus and invading optic nerve and extended posteriorly intracranially.

**Figure 2**



MRI axial revealed the presence of a lobulated mass with soft parts density in the Rt ethmoid sinus and invading the Orbital contents engulfing the optic nerve.

**Figure 4**



MRI showed intracranial extension of the mass.

- (1) Extracranial extensions of a meningioma with an intracranial origin (secondary)
- (2) Extracranial extensions of a meningioma arising in neural foramina (primary)
- (3) Ectopic, without any connection either to the foramen of a cranial nerve or to intracranial structures (primary)
- (4) Extracranial metastasis from an intracranial meningioma (secondary).

As regards histology, meningotheliomas are reported (i.e., lobule cells with irregular outline and nucleolus increased by visible roseate chromatin) besides more common transitional cells and psammoma bodies in its interior [2].

Transitional meningiomas are a very common type of such tumor (75% of cases), followed by syncytial and fibrous tumors (10% each), angioblastomas (5%), and sarcomas (<1%) [2].

Diagnosis is made with the examination of the pathological material. Such neoplasm, involving the nasal cavity or the paranasal sinus, is common and its development is slow. When surgically removed, prognosis of primary meningioma of the nasal cavity is favorable with a low rate of reported recurrences. Therefore, prognosis of patients with intracranial involvement is less favorable [5].

An additional therapy, such as radiotherapy, was less efficient, even for disease recurrence [1].

Swain *et al.* [7] have found three cases of paranasal sinus meningiomas, including the frontal, ethmoid, and sphenoid sinuses when revising a retrospective study in the past 25 years.

## Conclusion

Meningiomas of the sinonasal tract are uncommon. The clinical and radiologic criteria of such tumors are unspecified, and hence acute diagnosis requires

histopathological evaluation. Meningiomas of the sinonasal tract are histologically and immunophenotypically undistinguishable from intracranial ones. Comprehensive surgical resection in meningiomas of the sinonasal tract is usually well prognosticated [1]. Radiotherapy is used in unresectable malignant meningioma or recurrent meningioma for which surgery is not feasible [3,4].

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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