

Primary extracranial meningioma

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Primary [extracranial meningioma](#) is a rare type of tumor that originates from the [meninges](#), which are the protective membranes that surround the brain and spinal cord. [Meningiomas](#) are typically intracranial tumors, but in some rare cases, they can occur in extracranial locations, meaning they develop outside of the skull and spinal column. These tumors are also referred to as ectopic or extracranial meningiomas.

Here are some key points about primary extracranial meningiomas:

Location: Unlike intracranial meningiomas, which are found within the skull, primary extracranial meningiomas occur outside the cranial vault. They can develop in various extracranial sites, such as the scalp, orbit (eye socket), nasal cavity, paranasal sinuses, and neck.

Rarity: Extracranial meningiomas are extremely rare, accounting for a small fraction of all meningioma cases. The majority of meningiomas are intracranial.

Histology: These tumors exhibit similar histological features to intracranial meningiomas. They are typically classified based on their histological subtypes, such as meningothelial, fibrous, transitional, and psammomatous.

Clinical Presentation: The clinical presentation of primary extracranial meningiomas depends on their location. Patients may experience symptoms related to the compression of nearby structures. For example, a meningioma in the nasal cavity may lead to nasal obstruction and sinusitis, while an orbital meningioma may cause proptosis (bulging of the eye).

Diagnosis: Diagnosis is typically made through imaging studies such as CT scans or MRI scans. Biopsy and histopathological examination are required to confirm the diagnosis and classify the tumor.

Treatment: Treatment options for primary extracranial meningiomas include surgical resection, radiation therapy, and in some cases, adjuvant therapy. The choice of treatment depends on factors like tumor size, location, histology, and the patient's overall health.

Prognosis: The prognosis for extracranial meningiomas varies depending on factors such as tumor location and histological subtype. Generally, these tumors are considered less aggressive than certain intracranial meningiomas, and the prognosis can be favorable, especially with complete surgical removal.

It's important to note that primary extracranial meningiomas are distinct from metastatic meningiomas, which are extremely rare and involve the spread of intracranial meningioma cells to extracranial sites. Primary extracranial meningiomas are self-contained tumors that originate in extracranial tissues.

A female patient in her early 20s was found to have a mass on her first screening pelvic examination. Subsequent imaging followed by surgical resection was performed with the final diagnosis of a pelvic meningioma. Routine pelvic examinations in asymptomatic women may be more useful than merely screening for cervical cancer and sexually transmitted infections. Once detected, the differential diagnosis of a pelvic mass may include aetiologies outside of the gynecological organ system ¹⁾.

Extracranial meningiomas of the paranasal sinus are the rare tumors of unclear etiology. We report a case of primary extracranial meningioma arising from the roof of the sphenoid sinus. A 60-year-old female presented with 5-month history of right visual disturbance. Imaging studies showed a mass in the sphenoid sinus extending into posterior ethmoid sinus. The tumor was resected completely including the affected bone of anterior cranial base via the endonasal endoscopic approach. Pathological diagnosis was meningothelial meningioma. On the basis of imaging features and intraoperative findings, the tumor was diagnosed as primary extracranial meningioma of the sphenothmoid sinus. Recent advancement of skull base surgery technique allows removing this rare tumor completely and safely without complications such as cerebrospinal fluid leakage. This is the first report of the management of sphenothmoid sinus meningioma with the skull base surgery technique ²⁾

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Carlson K, Wegner L. Primary extracranial meningioma of the pelvis discovered on screening pelvic examination. BMJ Case Rep. 2023 Sep 25;16(9):e256988. doi: 10.1136/bcr-2023-256988. PMID: 37748813.

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Mori R, Cavallo LM, Guadagno E, Imperato A, Murayama Y, Cappabianca P. Extracranial Sphenothmoidal Sinus Meningioma: Case Report. Turk Neurosurg. 2014;24(5):788-792. doi: 10.5137/1019-5149.JTN.9452-13.0. PubMed PMID: 25269056.

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