Case Report

Atypical olfactory groove meningioma associated with uterine fibromatosis; case report

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Abstract

The concomitant presence of the olfactory groove meningioma with uterine fibrosis is very rare. Our report presents the case of a giant olfactory groove meningioma revealed after a uterine fibroma resection in a 44 years-old female, due to a generalized seizure 10 days after operation.

Cranial CT-scan identified the tumor as an olfactory groove meningioma. The tumor was operated with a macroscopically complete resection; the endothermal coagulation of the dura attachment was performed (Simpson II) with a good postoperative evolution. Laboratory results showed the presence of receptors for steroid hormones both in meningioma and uterine tumor, and the histopathological examination revealed an atypical meningioma with 17% proliferation markers.

Our findings suggest that even though meningiomas are benign tumors and a complete resection usually indicates a good prognosis, the association with uterine fibromatosis and the presence of high percentage of steroid receptors creates a higher risk to relapse, imposing therefore a good monitoring.

Keywords: olfactory groove meningioma, uterine fibromatosis, association

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Introduction

Olfactory groove meningiomas are intracranial tumors that develop from the arachnoid cells of the olfactory groove and crista galli. They appear more frequently during adulthood, especially in female (1-3).

Clinical picture includes signs and symptoms related to the compression of the neighboring anatomical structures: smell disorders which may evolve to anosmia, visual disorders, intracranial hypertension signs, compression of the frontal lobes followed by seizures, sphincter incontinence, memory, behavior and affectivity disorders. The Foster Kennedy syndrome (anosmia, optic atrophy in the ipsilateral eye and papilledema in the contralateral eye) is typical for such meningiomas (4, 5).

The CT scan shows the olfactory groove meningioma as a hyperdense, homogenous, densely enhancing mass, with a broad base along the dural insertion and adjacent edema in variable degrees extended into the cerebral white matter. Intratumoral calcifications (60-70 Hounsfield units) and hyperostosis are suggestive on the CT scan when viewed in bone window.

Histopathologically they can be benign tumors (meningothelial, fibroblastic, transitional, psammommatous, angiomatous, microcystic, secretory, clear cells, cordoid), semi-malignant tumors (atypical, papillary) and malignant (anaplastic).

Several immunohistochemical intra and extracellular markers were described for the atypical cerebral meningioma identification: vimentin (epithelial membrane antigen), but also progesterone receptors and other proliferation and prognosis markers (6, 7).

The neurosurgical treatment consists of the bifrontal/ unilateral craniotomy and tumor removal by internal debulking with sectioning of the insertion base for interrupting the tumor vascularization. Duroplasty is performed using a vascularized pericranial flap with the base on the supraorbital rim and hinged like a curtain over the frontal sinuses and the ethmoidal cells.

Case Report

**History and examination**

A 44-year-old female with total hysterectomy for a large volume, solid, abdominopelvic mass - 12/18 cm in axial plane (Figure 1), presented 10 hours postoperatively a seizure. Postcritically were noticed: divergent strabismus, right anisocoria, and bilateral Babinski sign.

![Figure 1](https://example.com/figure1.png)

**Figure 1.** Uterine fibrous tumor (12/18 cm) on abdominopelvic native CT- scan (a) and contrast-enhanced CT - scan (b).

The abdomino-pelvic native CT scan was normal postoperatively. The native and contrast cerebral CT (Figure 2a) shows an intra-axial tumoral lesion measuring 50/48/44 mm, with antero-basal location and intensely inhomogeneous structure with alternating areas of intense and mild contrast enhancement, digitiform perilesional edema in the bilateral frontal white matter, and mass effect on the anterior horns of the lateral ventricles predominantly on the right side.

The patient presented a favorable neurological evolution after anti-edematous treatment (diuretics, corticotherapy), and was transferred in the neurosurgical department.
Operative and postoperative course

The neurosurgical procedure started with a bifrontal, Souttar-type incision, followed by a bifrontal craniotomy. The dura was cut semicircular with a median pedicle, bilaterally.

The superior sagittal sinus was ligated and cut in its anterior extremity, and then the falk was cut following the sinus section line. Total macroscopic resection of a well-vascularized, white-yellowish, with low consistency, extracerebral tumor, inserted on the right olfactory groove, was done (Figure 3).

The dural insertion of the tumor was coagulated (grade II Simpson resection). The cranialization of the frontal sinuses was followed by their filling with autologous fat from the abdominal wall. The fat was covered with a pericranial vascularized graft. A subgaleal drain was left on place. The bone flap was replaced.

Microscopically, the tumor consists of meningothelial-like cells arranged in sheet-like syncytial placards, with extended area of necrosis, moderate pleomorphism of the nuclei and isolated calcifications, suggestive aspects for an atypical meningioma. The immunohistochemical examination (OMS -9539/1 criteria) confirmed the transformed cells type and their biological activity: cells diffuse positive vimentin, locally positive for the epithelial membrane antigen, 60% positive for the progesterone receptors. The proliferation and prognosis Ki-67 (MIB-1) index for the tumor was 17%.

The postoperative local and general evolution of the patient was favorable (Figure 2b).

Discussion

Olfactory groove meningiomas account for 4.8% of cerebral meningiomas and they develop in a neurological mute area so they reach a considerable size when diagnosed.

Atypical meningioma presents at least 3 out of next 4 histological parameters: hypercellularity, macronucleoli, small size cells, and the lack of architecture (8). According to these, our case is an atypical meningioma being confirmed immunohistochemically by intra and extracellular markers. The intermediate filaments from the meningotelial-like cells cytoplasm seem to correspond to vimentin immunostaining (1).

The presence of estrogen and progesterone receptors was confirmed in cerebral meningiomas (9, 10). These receptors are also present in the tumoral tissue from uterus and mammary glands and some
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Meningiomas are associated with breast cancer (11). The common features of these tumors are: they appear in females, in the 4th-5th decade of life and are worsened during pregnancy. They present common hormonal receptors, but in different ratio: the number of estrogentic hormone receptors is low in meningiomas and high in breast cancer while progesteronic receptors are high in both tumors. The proliferation rate of the cerebral meningioma is modulated by the female steroid hormones (12). In our case, the hormone receptors for progesterone were positive in 60% of cells. There were not identified estrogen receptors in the meningioma. An increased index of the Ki-67 proliferation factor reflects an aggressive behavior of the tumor and an increased risk of recurrence. In our case, the Ki-67 proliferation factor index was 17% (13-15).

Conclusions

Olfactory groove atypical meningioma is a rare entity with an uncertain behavior, benign or malignant, with an increased rate of proliferation and a relatively increased rate of recurrence comparative with other meningiomas.

Surgical treatment remains the best option, consisting of a radical tumor removal. Imagistic monitoring is necessary due to the risk of relapse.

References


