Atypical olfactory groove meningioma associated with uterine fibromatosis

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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 on, a macroscopically complete resection with endo-thermal 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 and imposes a closed monitoring.
**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, 5).

Clinical picture includes signs and symptoms related to the compression of the neighbouring 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, behaviour and affectivity disorders. The Foster Kennedy syndrome (anosmia, optic atrophy in the ipsilateral eye and papilledema in the contralateral eye) is typical for these meningiomas (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, psamommatous, angiomatous, microcystic, secretory, clear cells, cordoid), semi malignant tumours (atypical, papillary) and malignant (anaplastic).

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

The neurosurgical treatment consists of the bifrontal/unilateral craniotomy and tumour removal by internal debulking with sectioning of the insertion base for interrupting the tumour vascularisation. Duroplasty is performed using a vascularised 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 (Fig.1), presented 10 hours postoperatively a seizure. Post critically were noticed: divergent strabismus, right anisocoria, bilateral Babinski sign.

Fig. 1 Uterine fibromatous tumour (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 (Fig.2 A) shows an intraaxial tumoral lesion measuring 50/48/44 mm, with anterior-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 had a favorable neurological outcome after anti-oedematous 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 craniectomy. The dura was cut semi-circular with a median pedicle, bilaterally.

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

Fig. 3 Macropopical aspect of the removed olfactory groove atypical meningioma

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 peri-cranial vascularised graft. A sub-galeal drain was left on place. The bone flap was replaced.

Microscopically, the tumour consists of meningo-telial-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 immunohistochemically 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 outcome of the patient was favorable, local and general (Fig 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, the lack of architecture (8). According to these, our case is an atypical meningioma and is immunohistochemically confirmed by intra and extracellular markers. The intermediate filaments from the meningo-telial-like cells cytoplasm seem to correspond to vimentin immunostaining (1).

The presence of estrogen and progesterone receptors was confirmed in cerebral meningiomas. 6,7). These receptors are also present in the tumoral tissue from uterus and mammary glands and some meningiomas are associated with breast cancer (13). The common features of these tumours 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 estrogenic 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 (4). 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 behaviour of the tumour and an increased risk of recurrence. In our case, the Ki-67 proliferation factor index was 17%.

**Conclusion**

Olfactory groove atypical meningioma is a rare entity with an uncertain behaviour, benign or malignant, with an increased rate of proliferation and a relatively increased rate of recurrence comparative with other meningiomas (11).

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

**References**


