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Case Report

Unusual coexistence of an epidermoid cyst with an atypical meningioma: Case report and review of the literature

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Abstract

Background: Coexistence of multiple primary intracranial tumors of different cell types has rarely been documented; the association of a meningioma and a glioma has been reported as the most common combination. Hereby, we report an unusual case of a temporal epidermoid cyst coexisting with an atypical meningioma.

Case Presentation: A 37-year-old male presented with progressive symptoms of raised intracranial progression with progressive loss of vision without any neurological deficit. On admission, magnetic resonance imaging (MRI) revealed a right frontal lesion appearing hypointense T1, hyperintense T2 slightly enhanced after gadolinium and a second right temporal, isointense T1, hyperintense T2 non-enhancing lesion. A right frontotemporal craniotomy was performed that revealed two distinct lesions: The whitish temporal lesion with the pearl appearance reminding of an epidermoid cyst, the second lesion was extraaxial fibrous lesion arising from the falx. Pathology confirmed an atypical meningioma WHO Grade II and an epidermoid cyst.

Conclusion: The simultaneous occurrence of primary intracranial tumors of different cell types is rare. Epidermoid cysts are slow growing lesions believed to arise from inclusion of ectodermal elements during neural tube closure, while meningiomas arise from arachnoidal cells; their association has rarely been reported previously.

Key Words: Epidermoid cyst, meningioma, multiple tumors

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INTRODUCTION

Epidermoid cysts are benign, slow growing extraaxial tumors that account for about 1% of all intracranial tumors;[3] they arise from hamartomatous misplacement of embryonic ectoderm during ontogenesis. Meningiomas, on the other hand, arise from arachnoidal cells.[4] Except for cases associated with neurofibromatosis type 2, multiple primary intracranial tumors of different cell types are relatively rare. The most frequent combination of multiple tumors is a meningioma with a glioma.[8,20]

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Association of a meningioma and an epidermoid cyst has rarely been reported before. We present an unusual case of a temporal epidermoid cyst coexisting with an intracranial meningioma and review the possible pathogenesis of association of those lesions.

CASE REPORT

We report the case of a 37-year-old patient who presented with symptoms of raised intracranial pressure with progressive loss of sight. On admission, patient was conscious with no motor deficit, and fundus revealed bilateral optic atrophy. Magnetic resonance imaging (MRI) revealed an extra-axial right frontal lesion, hypointense T1, hyperintense T2 slightly enhanced after gadolinium; a second right temporal lesion isointense T1, hyperintense T2 non-enhancing was seen [Figure 1]. A right frontotemporal craniotomy was performed that revealed two distinct lesions: The temporal whitish lesion with the pearl appearance reminding of an epidermoid cyst and the second was extraaxial, fibrous arising from the falx reminding of a meningioma. Pathology confirmed an atypical Grade II meningioma and epidermoid cyst [Figure 2]. The patient recovered well following surgery and had no neurological deficit except his previous optic atrophy. Postoperative computer tomography (CT) scan showed total removal of both lesions [Figure 3].

DISCUSSION

Intracranial epidermoid cysts account for 0.2–1%[3] of all brain tumors. They are thought to derive from ectodermal cell inclusions occurring during closure of the neural tube; embryologically, epithelial components of the cyst wall develop between the 3rd and 5th weeks of embryonic life; posttraumatic acquired examples have been reported.[25] Epidermoid cysts occur mostly at the cerebellopontine angles (40–50%) and parasellar regions, insinuating between brain structures. They are usually benign and slow growing lesions, although rare malignant transformation into squamous cell carcinoma has been documented in the literature.[1] Meningiomas, on the other hand, arise from the arachnoidal cap cells of arachnoid villi in meninges; they are classified as benign, atypical, and anaplastic and account for up to 24–30% of all primary intracranial neoplasms, the majority being benign.[4]

Simultaneous occurrences of primary intracranial tumors of different cell types have rarely been documented. There are few cases in the literature, the coexistence of a meningioma and a glioma being the most reported combination, followed by a meningioma with a vestibular schwannoma and a meningioma with a pituitary adenoma.[3,7,13-19,21,25] The recent advance in neuroimaging for the diagnosis of brain tumors has resulted in more additional reports of multiple primary brain tumors, with multiple other cases of coexistence of pituitary adenoma and temporal lobe glioma,[9] prolactinoma

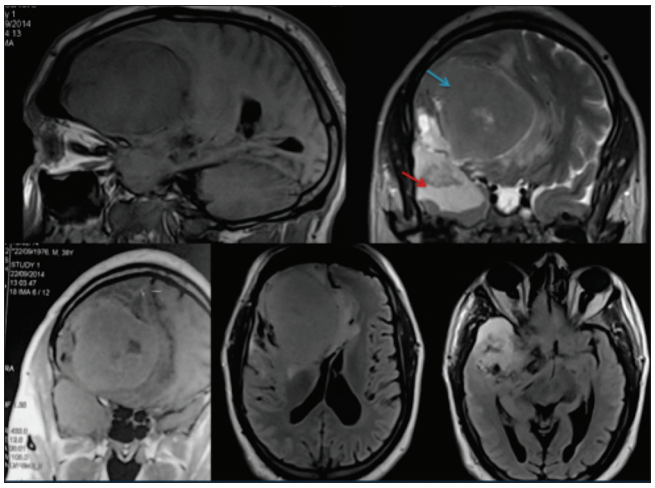

Figure 1: A right extra-axial, frontal lesion hypointense T1, hyperintense T2 slightly enhanced after gadolinium; a second right temporal lesion isointense T1, hyperintense T2 non-enhancing seen

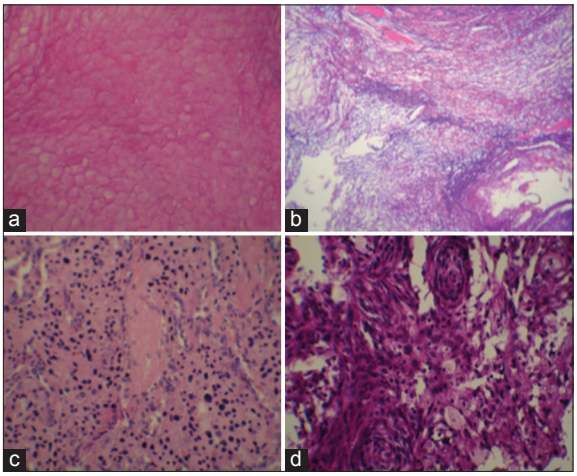

Figure 2: (a and b) Stratified epithelium containing keratin and cholesterol reminding of an epidermoid cyst. (c and d) Clear small cells with prominent nucleoli, cell change, infiltration, and necrosis suggesting of an atypical meningioma

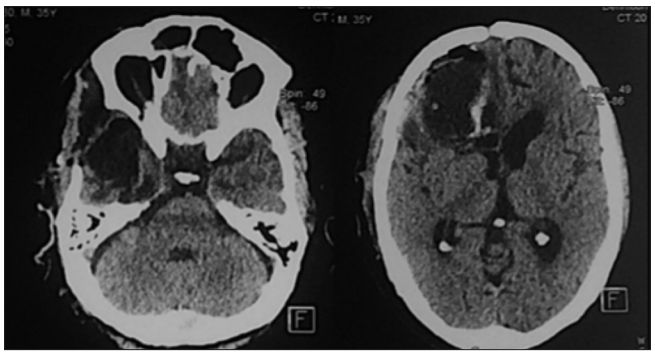

Figure 3: Control computer tomography (CT) scan showing total removal of the two lesions

and craniopharyngioma,[5] epidermoid cyst tumor, and pituitary adenoma[11] described. Co-occurrence of meningioma and an epidermoid cyst has barely been reported previously. In 1979, Frazer and Victoratos[6] have been the first to report a case of an epidermoid cyst and meningioma sequentially developing in the aftermath of cranial radiotherapy and in 1999, Kumar *et al*.[12] reported an epidermoid cyst coexisting with a subcutaneous meningioma.

Much speculation remains regarding the origin of multiple tumors of different histological type. Some authors support the theory of locally acting oncogenic factors and others believe it would just be a pure coincidence.[24] A number of genetic factors may also be involved. Some of these genetic factors are well recognized, for example, Li–Fraumeni syndrome that greatly increases the risk of developing several types of tumor, particularly in children and young adults, but there may be many unrecognized genetic associations as well. Another theory is that a meningioma or a glioma can stimulate the adjacent brain parenchyma or arachnoid cells into neoplastic proliferation.[23,24] In their case, Suzuki *et al*.[23] found high levels of expression of epidermal growth factor (EGF) receptor in tumor cells of a meningioma adjacent to a glioblastoma, and suggested that "the former might have exerted a growth promoting effect on the latter."

Epidermoid cysts are known to possess an arachnoid cap, which is a cerebrospinal fluid (CSF) collection surrounding the tumor.[22,24] Based on this knowledge, perhaps, the presence of a meningioma and an epidermoid cyst could be the result of irritants or oncogenic factors leaking into the CSF leading to the stimulation of tumor growth in susceptible areas. Since meningiomas are also known to both depend on and actively secrete EGF, paracrine stimulation might have played a role in initiating the development of the epidermoid cyst in our case as well.[10] The close proximity of the two tumors in our patients suggests that such local causes are probably involved, but for tumors located in different lobes or hemispheres, this remains unclear.

Treatment approach can be a debate topic. In the literature, the glial tumor usually had been the first target.[2,5,8] Our patient's complaints were related with the meningioma, the epidermoid cyst was totally incidental. In several other reported cases, one tumor was removed operatively and the second tumor was diagnosed at autopsy; in the case reported by Alexander,[2] a frontal meningioma was removed operatively and at autopsy, a tumor designated as a "spongioblastoma ependymale" was found in the roof of the left lateral ventricle. In 1950, Gass and van Wagenen[7] reported the first successful removal of adjacent tumors when a small meningioma and an underlying oligodendroglioma were removed in the same operation.

Nowadays, both CT and MRI are useful for intra- and extra-axial brain tumors. Diagnosis from CT scans can sometimes lead to misinterpretation where low-density areas may represent edema, ischemic change, or a low- to medium-grade astrocytoma.[20] MRI offers better definition than CT, particularly with the use of marked T2-weighted images and enhancement with gadolinium. However, differentiating peri-meningioma edema from cystic lesions can still be a problem with enhanced MRI scans.[20,21] In our patient, the temporal epidermoid cyst was misinterpreted as perilesional edema lesion on the original MRI.

Frequently, operative management is undertaken for what is assumed to be a single tumor, but the surgeon should be alerted to the existence of a second tumor by the presence of edema not consistent with the first tumor or when the first tumor does not correlate with the preoperative radiographic findings.

CONCLUSION

Multiple primary intracranial tumors of different histological origins are rare; their possibility ought to be considered when symptoms and signs, intraoperative findings, or unexpected postoperative events do not correlate with radiological findings. Their treatment approach should be decided depending on the prognosis of the lesions and their localization in brain.

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Conflicts of interest
There are no conflicts of interest.

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