A Case of Multiple Primary Tumors of the Anterior Skull Base

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ABSTRACT

We report a case of synchronous olfactory bulb meningioma and undifferentiated carcinoma of the nose and paranasal sinuses that involved and destroyed the anterior skull base and mimicked intracranial invasion by a carcinoma. The heterogeneity of tissue types in the skull base gives rise to a diverse variety of benign and malignant neoplasms which have totally different prognoses. Synchronous development of benign and malignant primary tumors both originating from and involving the skull base at the same location is very rare and may cause confusion for both the skull base surgeon and neuroradiologist.

KEYWORDS: Meningioma, undifferentiated carcinoma, skull base, paranasal sinuses

The heterogeneity of tissue types in the skull base gives rise to number of benign and malignant neoplasms which have widely variable prognoses.¹ Synchronous development of benign and malignant primary tumors that both arise from and involve the skull base at the same location is very rare and can lead to a challenging preoperative diagnostic dilemma. A review of the literature shows several cases of synchronous development of meningiomas and other distant primary malignant tumors. More than 90% of these patients have breast or lung cancers. There are even reports of tumor-to-tumor metasta-

ses from these malignancies to the intracranial meningiomas.²⁻⁴ The association of breast cancer and meningioma suggests a possible hormonal relationship between the two pathological entities.³

CASE REPORT

A 66-year-old female presented with a 3-month history of progressive right-sided nasal obstruction and anosmia. Endoscopic examination of the nose

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and paranasal sinuses revealed a soft-tissue mass in her right nasal cavity that extended into the ethmoid sinus and anterior skull base. A computed tomography scan showed that the tumor was at least 3.5 cm in diameter and had destroyed the middle turbinate and nasal septum (Fig. 1A). There was complete opacification of the ethmoidal sinuses and frontal recess together with erosion of the right cribriform plate that suggested possible intracranial tumor spread. Magnetic resonance imaging (MRI) confirmed intradural extension of the tumor into the anterior cranial fossa in the region of the olfactory groove. The intracranial part measured 1.5 cm (Figs. 1B–D). No cervical lymphadenopathy

could be detected by either MRI or ultrasound examination. Biopsy of the intranasal tumor confirmed the diagnosis of an undifferentiated carcinoma.

A combined craniofacial resection through a right lateral rhinotomy and a right frontolateral craniotomy was undertaken. Because of its highly malignant nature, a radical resection of the tumor using image guidance was performed. The resected specimen included the middle turbinate, the infiltrated part of the septum, the medial wall of the maxillary sinus (preserving the nasolacrimal duct), medial part of the bony orbital floor, lamina papyracea, and ethmoid and sphenoid sinuses.

Figure 1 (A) Preoperative computed tomography scan shows bony destruction of the middle turbinate, nasal septum, and cribriform plate (arrow). (B) Preoperative axial MRI T1-weighted image shows the intranasal part of the tumor (arrows). (C) Preoperative coronal MRI T1-weighted image shows the intracranial and intranasal components of the tumor (arrows). (D) Preoperative axial MRI T1-weighted image shows the intracranial component of the tumor (arrows). MRI, magnetic resonance imaging.

Figure 2 (A) Coronal computed tomography scan shows the postoperative result. (B) Coronal MRI T1-weighted image shows the postoperative result. MRI, magnetic resonance imaging.

Intraoperative inspection showed tumor extension up to the skull base and obvious bony destruction at the anterior part of the cribriform plate. Intraoperative frozen section control of the resection margins confirmed the complete removal of the intranasal part of the tumor. Exposure of the intracranial part through the right frontolateral craniotomy gave adequate access to the lobulated intradural component of the tumor in the region of the olfactory groove. It was well encapsulated and sharply defined from the surrounding brain tissue, but was adherent to the underlying dura. The tumor was completely removed together with the underlying dura, and the suspicious areas of eroded bone around the cribriform plate were drilled out extensively. The dura defect was reconstructed using a vascularized pericranial galeal-periosteal flap to prevent cerebrospinal fluid leakage (Figs. 2A, B).

The patient's postoperative course was uncomplicated and she recovered quickly without acquiring any new neurological deficits. No cerebrospinal fluid leakage developed.

Histological examination of the intranasal tumor confirmed the diagnosis of an invasive undifferentiated carcinoma (Fig. 3). Surprisingly, histological examination of the intradural part revealed a benign olfactory groove meningioma as a second primary tumor, histologically unrelated to the intranasal finding (Fig. 4). There was no evidence of

carcinomatous infiltration of the dura. The patient received a course of adjuvant radiotherapy to the head and neck. There has been no recurrence after 2.5 years of regular follow-up. The cosmetic result of the operation has been very satisfactory.

DISCUSSION

Malignant sinonasal tumors usually present with nasal obstruction, epistaxis, purulent rhinorrhea,

Figure 3 Undifferentiated carcinoma composed of solid sheets of neoplastic epithelial cells with increased nuclear/cytoplasmic ratio, pleomorphic nuclei with prominent nucleoli, and numerous mitotic figures. Paraffin section, hematoxylin and eosin.

Figure 4 Meningothelial meningioma, composed of lobuli of monomorphic cells, and a psammoma body. Paraffin section, hematoxylin and eosin.

postnasal drip, and hyposmia or anosmia. Adenocarcinomas predominate with squamous cell carcinoma and adenoid cystic carcinoma being less frequent. Undifferentiated carcinomas are rare.⁵ Local recurrence and invasion to the skull base are two of the common features of all malignant sinonasal tumors. The cribriform plate is a potential route of extension into the anterior cranial fossa. MRI is helpful to evaluate intracranial extension, and findings that are usually suggestive of dural invasion include nodular, irregular enhancement of the dura, evidence of dural thickness (more than 5 mm), pial and parenchymal enhancement, or brain edema.⁶

Olfactory groove meningioma is also a rare clinical entity. The principal symptoms associated with this tumor are anosmia and headache. This sometimes prompts referral to an ENT specialist. This tumor appears in the midline of the anterior cranial fossa along the dura of the cribriform plate and planum sphenoidale. It is often associated with bony changes such as hyperostosis or thinning and even destruction of the underlying bone, which in

15% of the cases leads to direct extension of the tumor into the neighboring ethmoid and sphenoid sinuses. The recurrence rate of olfactory groove meningiomas ranges from 5 to 41% and depends on the extent of resection and duration of follow-up. Radical tumor removal, including the dural attachments and any involved bone, is the best way to reduce any chances of recurrence. The risk of cerebrospinal fluid leakage after radical resection can be reduced by careful reconstruction of the anterior cranial fossa.⁷

To date, no etiological relationship has been found between these two tumors. A review of the literature showed that no other cases had been reported. It may be a rare coincidence, but the report of such cases may in the future enable us to find a potential relationship between these different pathologic entities involving the skull base.

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