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（收稿日期：2017-03-14）

·临床医学图像·

嗅神经母细胞瘤

doi：10.3969/j.issn.1672-6731.2017.05.015

Olfactory neuroblastoma

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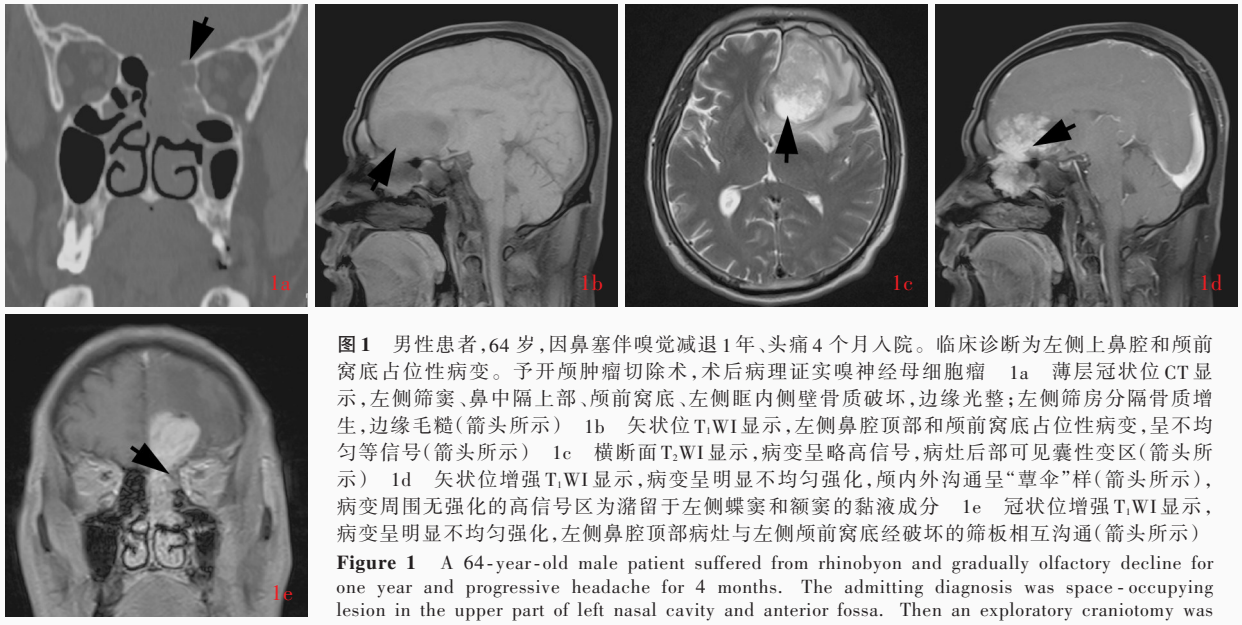

图1 男性患者，64岁，因鼻塞伴嗅觉减退1年、头痛4个月入院。临床诊断为左侧上鼻腔和颅前窝底占位性病变。予开颅肿瘤切除术，术后病理证实嗅神经母细胞瘤 1a 薄层冠状位CT显示，左侧筛窦、鼻中隔上部、颅前窝底、左侧眶内侧壁骨质破坏，边缘光整；左侧筛房分隔骨质增生，边缘毛糙（箭头所示） 1b 矢状位T_1WI显示，左侧鼻腔顶部和颅前窝底占位性病变，呈不均匀等信号（箭头所示） 1c 横断面T_2WI显示，病变呈略高信号，病灶后部可见囊性变区（箭头所示） 1d 矢状位增强T_1WI显示，病变呈明显不均匀强化，颅内外沟通呈“蕈伞”样（箭头所示），病变周围无强化的高信号区为潴留于左侧蝶窦和额窦的黏液成分 1e 冠状位增强T_1WI显示，病变呈明显不均匀强化，左侧鼻腔顶部病灶与左侧颅前窝底经破坏的筛板相互沟通（箭头所示）

Figure 1 A 64-year-old male patient suffered from rhinobyon and gradually olfactory decline for one year and progressive headache for 4 months. The admitting diagnosis was space-occupying lesion in the upper part of left nasal cavity and anterior fossa. Then an exploratory craniotomy was performed and postoperative pathological diagnosis was olfactory neuroblastoma. Thin coronal CT revealed well-defined bony destruction involving left ethmoidal sinus, upper body of nasal septum, base of anterior fossa and medial wall of left orbit. There existed hyperostosis of the separate bony wall of left ethmoidal cell with coarse margin (arrow indicates, Panel 1a). Sagittal T_1WI showed heterogeneous isointense space-occupying lesion located in the upper part of left nasal cavity and anterior fossa (arrow indicates, Panel 1b). Axial T_2WI showed slightly high-intensity lesion with cystic degeneration in the rear part (arrow indicates, Panel 1c). Sagittal enhanced T_1WI showed markedly heterogeneous enhancement both in extracranial and intracranial part, which looked like a "mushroom" (arrow indicates). The high-intensity areas with no contrast adjacent to enhanced tumor were myxoid components retention in left sphenoid sinus and frontal sinus (Panel 1d). Coronal enhanced T_1WI showed markedly heterogeneous enhanced lesion. The lesion in upper part of left nasal cavity was connected with the base of left anterior fossa through damaged lamina (arrow indicates, Panel 1e).

嗅神经母细胞瘤（ONB）是少见的源于鼻腔嗅觉受体细胞的神经外胚层肿瘤，发生部位与嗅黏膜分布区一致，包括上鼻甲、鼻中隔上部、鼻根部和筛孔等鼻腔顶部和近中鼻甲外侧壁。CT薄层扫描和多平面重建可见细微骨质改变，对病变进展有明确提示意义。肿瘤体积较小时仅局限于鼻腔（改良Kadish分期A期），呈较均匀的等或稍低密度影；体积较大时密度不均匀，其内有小囊性变和钙化，侵及副鼻窦（B期）以及筛板、眼眶和颅前窝底（C期），亦可穿透筛板累及颅内；出现颈部淋巴结转移或远处转移（D期）易漏诊。鼻中隔上部、颅前窝底骨质和眼眶内壁易出现骨质破坏。由于肿瘤生长缓慢且呈膨胀性生长，骨质破坏边缘较光整；部分患者因长期刺激而出现邻近骨质增生或骨质破坏与增生并存征象，是明确诊断的重要标记（图1a）。MRI在显示病变侵及范围方面更准确，肿瘤体积较小时信号较均匀，T_1WI呈等或稍低信号（图1b）、T_2WI呈等或稍高信号，体积较大时因囊性变和少量钙化致信号不均匀；侵及颅内时，肿瘤边缘可见更高信号的囊性变区（图1c）。DWI呈稍高或高信号。增强扫描多呈中度或明显不均匀强化，病灶内或边缘可见不同程度线状、环状或花环状强化（图1d），与肿瘤内纤维网状结构致细胞团块样排列和病灶内网状增生的血管袢有关。病灶内囊性变和钙化呈非强化改变。增强扫描有助于区分肿瘤与周围窦腔内黏液组织。肿瘤沿嗅神经向上侵犯颅内，形成颅内外沟通，呈现以筛板为中心的“哑铃”样或“蕈伞”样改变（图1e）。

（天津市环湖医院神经放射科韩彤供稿）