Olfactory neuroblastoma
A frontal lobe disorder and a runny nose

A 56-year-old woman presented with progressive apathy, expressive aphasia, urinary incontinence, and a subtle right hemiparesis. She had a recent history of clear rhinorrhea and frequent nosebleeds. CT and MRI scan revealed a large mass in the left nasal cavity extending into the frontal lobe (figure 1). The diagnosis of olfactory neuroblastoma was confirmed by transnasal biopsy (figure 2). A combined treatment of chemotherapy and radiotherapy was suggested. Before treatment could be initiated, the patient died of a major hemorrhage in the tumor.

Olfactory neuroblastoma is a rare (0.4 per million per year) neural crest tumor thought to arise from the olfactory neuroepithelium. Treatment generally consists of resection and radiotherapy with or without chemotherapy.1,2

Figure 1  Coronal CT and sagittal MRI

(A) Coronal CT scan shows a mass in the left nasal cavity, paranasal sinuses, and frontal lobe with destruction of the cribriform plate and medial orbital wall. There is vasogenic edema and subfalcine herniation. (B) T1-weighted contrast-enhanced MRI shows a dumbbell-shaped enhancing mass in the left frontal lobe, the nasal cavity, and the sphenoid sinus.

Figure 2  Pathology

(A) Pathologic examination shows small round blue cells in a diffuse and lobular arrangement with prominent areas of necrosis (hematoxylin & eosin stain, ×5). (B) Neoplastic cells show nuclear pleomorphism and marked mitotic activity (hematoxylin & eosin stain, ×40). (C) Immunohistochemistry is positive for neuroendocrine markers, with heterogeneous expression of synaptophysin (C).
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