

A Case Report of Mixed Olfactory Neuroblastoma — Neuroblastoma and Adenocarcinoma —

Yukihiko SATO, Toru HANDA, Yozo ORITA,*
Mitsuyoshi HIROKAWA** and Chotatsu TSUKAYAMA***

*Department of Otolaryngology,

**Department of Pathology

Kawasaki Medical School, Kurashiki 701-01, Japan

***Kurashiki Central Hospital, Kurashiki 710, Japan

Accepted for publication on May 29, 1997

ABSTRACT. We report a case of mixed olfactory neuroblastoma with two histologically different appearances, neuroblastoma and adenocarcinoma. The patient was a 30-year-old-man with a left submandibular mass. The submandibular mass was histologically diagnosed as an adenocarcinoma. From histological and immunohistochemical studies of the resected masses from the nasal cavity and the neck, we made a diagnosis of mixed olfactory neuroblastoma. Herein we discuss some of the diagnostic procedures for olfactory neuroblastoma and describe the importance of immunohistochemical staining.

Key words: mixed olfactory neuroblastoma — adenocarcinoma —
neuroblastoma

Olfactory neuroblastoma is an uncommon tumor that is considered to arise from the olfactory epithelium. Over 200 cases have been described in the medical literature, since the first case was reported in 1924.^{1,2)} Histologically, the tumor is characterized by organoid masses of primitive neurocytoblasts with neural fibrils, but no ganglion cells. A very rare olfactory neuroblastoma contains areas of divergent differentiation such as adenocarcinoma was reported in 1984.³⁾

We encountered a case of mixed olfactory neuroblastoma which was initially diagnosed as adenocarcinoma from a biopsy specimen of the nasal cavity. We report a case of a rare histological type of olfactory neuroblastoma and discuss the procedures involved in diagnosis.

CASE REPORT

A 30-year-old man presented with a left submandibular mass. Histologically, diagnosis of metastatic adenocarcinoma was made from biopsy specimens. Five months later, a left nasal obstruction and hyposmia appeared and a left nasal mass was noted (figure 1). A plain X-ray demonstrated opacity in the left maxillary and ethmoid sinuses (Figure 2). Computerized tomography (CT) revealed a mass in the left nasal cavity. The mass occupied the ethmoid sinus without bone destruction (Figure 3). A chest X-ray and other physical examination disclosed no particular findings. A biopsy of the

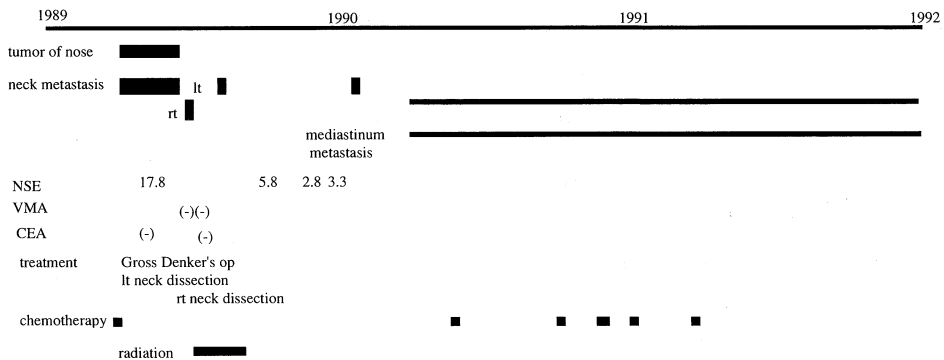


Fig 1. Clinical course of the present case

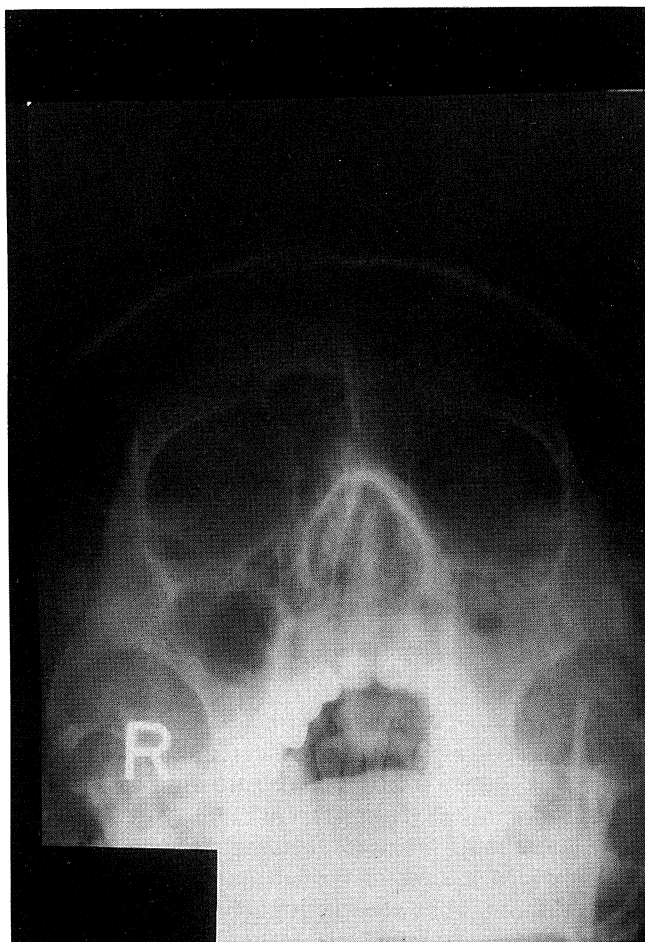


Fig 2. Radiograph of the head. An opacity of the left maxillary and ethmoid sinuses was seen.

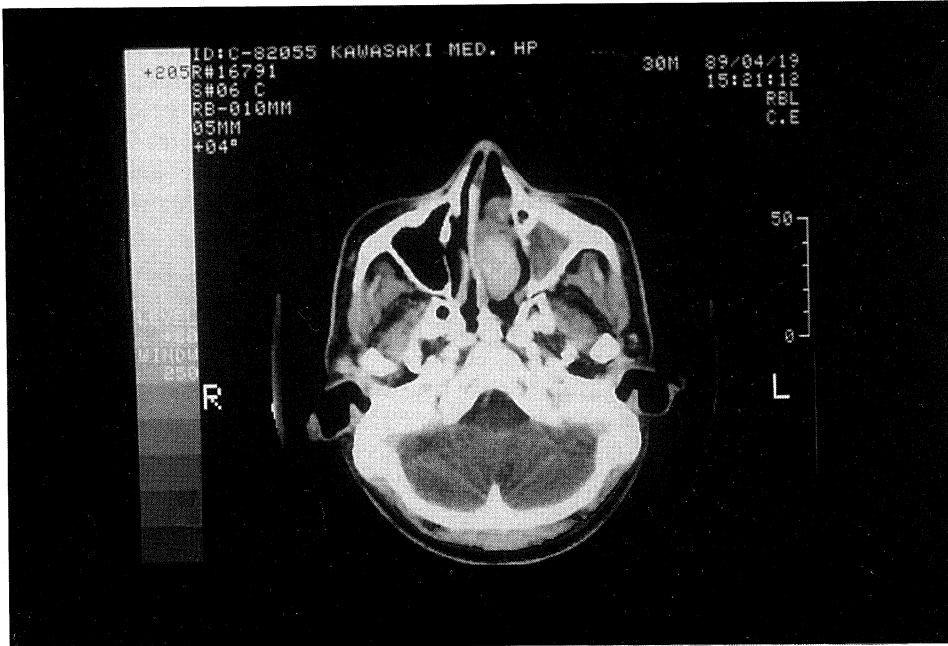


Fig 3. CT of the head. A mass lesion of the left nasal cavity was noted. Radiograph of the head. An opacity of the left maxillary and ethmoid sinuses was seen.

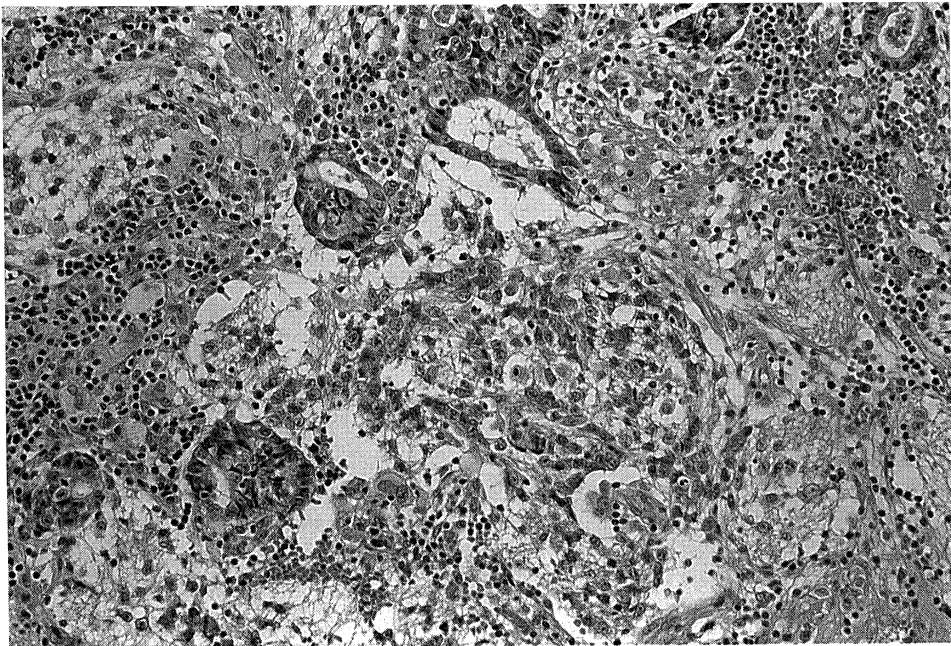


Fig 4. Histological appearance of the tumor. The tumor was composed of both neuroblastoma and adenocarcinoma. (H & E stain, X200)

nasal mass was performed and the histological diagnosis was adenocarcinoma. Methotrexate, nimustatine and therarubicin were intravenously administered before surgical resection. All of the visible tumor was surgically removed (Gross Denker's operation and right neck dissection). Macroscopically, the tumor existed in the nasal cavity and a neck metastasis existed in the right neck. The specimens showed both simple olfactory neuroblastoma and mixed olfactory neuroblastoma. No tumor existed in the maxillary and ethmoid sinuses. Histologically, the tumor consisted of tumor cells showing small round nuclei and indistinct cytoplasmic borders in a neurofibrillary background consistent with neuroblastoma. In some areas, there were also gland-forming tumor cells, indicating adenocarcinoma (Figure 4). In electromicroscopic observation, cytoplasmic processes containing membrane-bound granules are usually seen but typical feature was not seen. Immunohistochemically, the neuroblastoma cells were positive for neuron-specific enolase (NSE) and neurofilaments, and the adenocarcinoma cells were positive for carcinoemmbryonic antigen (CEA). From those results a definite diagnosis of mixed olfactory neuroblastoma was made.

Radiation therapy of the nasal cavity and neck metastasis was performed after resection. Three months later, metastasis to the neck occurred and complete surgical excision was performed. The resected mass indicated neuroblastoma.

A year later, however, metastasis to the mediastinum was found. Three years after the left mandibular mass appeared, the patient died of respiratory failure.

DISCUSSION

Olfactory neuroblastoma occurs in a broad age range, with bimodal age distribution having peaks at ages 11-20 years and 51-60 years.⁴⁾ There is no sexual predilection.²⁾ Presenting symptoms are usually nasal obstruction or epistaxis. In our case report, nasal findings were typical of olfactory neuroblastoma, but the histology was unusual. In our case, the olfactory neuroblastoma contained an adenocarcinoma component. Mixed olfactory neuroblastoma with adenocarcinoma was first reported by Douglas *et al* in 1984.³⁾ We have never seen a rare olfactory neuroblastoma contains adenocarcinoma till 1997. It is said that the adenocarcinoma component may represent the participation of neuroectodermal supporting cells from the olfactory epithelium in the tumor process. The prognostic value of this subtype has not been clarified.

Immunohistochemical stainings are very useful in differentiating olfactory neuroblastoma from other tumors. Furthermore, we were able to diagnose mixed olfactory neuroblastoma, neuroblastoma and adenocarcinoma. A majority of olfactory neuroblastomas are immunopositive for NSE, neurofilaments, S100 protein, chromogranin and Leu-7. NSE positivity is diffuse and commonly intense and acts as a preliminary neural tissue screener. Some cases reveal a positive keratin reaction. Glandular structures are immunopositive for CEA. A definite diagnosis in our case was made after immunohistochemical studies and its importance was reconfirmed.

REFERENCES

- 1) Berger L, Luc R, Richard J: L'esthesioneuroepitheliome, Olfactit. Bull. Ass. Franc Cancer **13**: 410-420, 1924
- 2) Baker DC, Perzin KH, Conley J: Olfactory neuroblastoma. Otolarygol Head Neck Surg **87**: 279-283, 1979
- 3) Douglas C Miller, Max L Goodman, Ben Z Pilch, Shan RongShi, G Richard Dickersin, Howard Halpern, Charles M Norris Jr: Mixed olfactory neuroblastoma and carcinoma. Cancer **54**: 2019-2028, 1984
- 4) Elkon D, Hightower SI, Lim ML, Cantrell RW, Constable WC: Esthesioneuroblastoma. Cancer **44**: 1087-1094, 1979
- 5) Banerjee-AK, Sharma-BS, Vashista RK, Kak Vk Intracranial olfactory neuroblastoma: Evidence for olfactory epithelial origin. J Clin Pathol **45**: 299-302, 1992
- 6) Frierson HF Jr, Ross GW, Mills Se, Frankfurter A Olfactory neuroblastoma. A dditional immunohistochemical characterization. Am J Clin Pathol **94**: 547-53, 1991