

Olfactory Neuroblastoma with Facial Metastasis

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Key words: Olfactory neuroblastoma, metastasis, face

Olfactory neuroblastoma (ON) is an uncommon malignant tumor. It arises from the olfactory epithelium, which is found in the cribriform region, the upper third of the nasal septum, and the superior and supreme nasal turbinates (1). This tumor was first described by Berger and Couthard (2) in 1924. ON shows a wide range of age distribution (3 to 79 years), the median age about 50 years. The clinical course is characterized by local aggressive and less commonly distant metastases (3,4). The most commonly involved sites are the cervical lymph nodes, but the lungs, long bones and pelvis are the other reported sites. There are isolated reports of metastasis to the scalp, orbit, aorta, spleen, liver, adrenal gland and ovary. Surgery combined with radiation therapy is the preferred treatment (5).

Case report

A 45-year-old woman was admitted to Atatürk University Research Hospital because of recurrent epistaxis, left facial swelling and nasal obstruction. Physical examination revealed a large granular mass confined to the left nasal cavity and a left facial mass. Grossly, the tumor was a polypoid mass, reddish-gray, soft and highly vascular. The mean diameter was 1.5 cm. Microscopically, the tumor was composed of uniform small cells with round nuclei, scanty cytoplasm, indistinct nuclear membrane, and a prominent fibrillary or reticular background. Hemorrhage and necrosis were seen (Figure 1). Immunocytochemically, tumor cells were marked by antineuron specific enolase antibodies (Figure 2). Anti-S-100 antibodies lightly marked the fusiform cells preferentially located at the periphery of tumor sheets, underlining cords; other cells were labelled by this antibody but were fewer in number and found scattered within the tumor sheets. Plain cranial roentgenograms and a computed tomography (CT) scan showed a mass occupying both nasal cavities with bilateral extension to maxillary and sphenoidal sinuse and medial wall of the orbits (Figure 3). Magnetic resonance imaging revealed the tumor extension to the anterior cranial fossa and epidural space recurrent epistaxis, left facial swollen condition, and nasal obstruction. Physical examination revealed a large granular mass confined to the left nasal cavity and left facial mass.

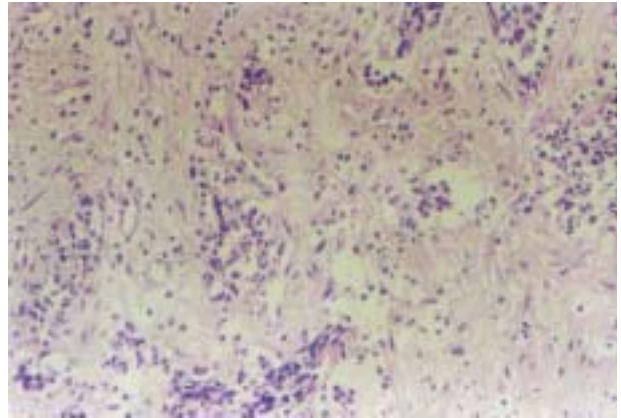


Figure 1. Olfactory neuroblastom exhibits discrete nests of small cells with bland cytologic feature. (H&E x 100)

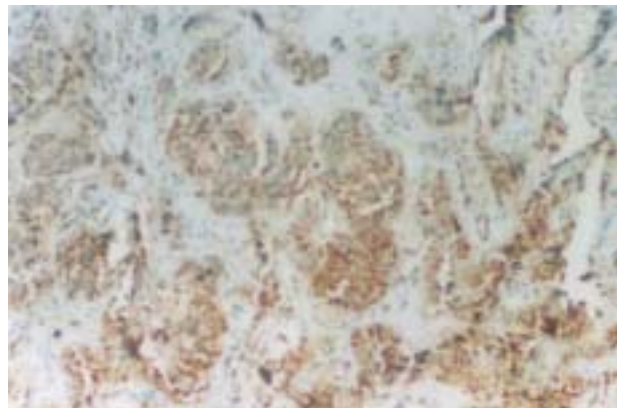


Figure 2. Positive immunostaining in tumor cells (antineuron-specific enolase). (Avidin–biotin-peroxidase complex x 200).

Discussion

The diagnosis of neuroblastoma by light microscopy depends on identification of dendritic processes, associated with the relatively uniform, small nuclei of the neuroblasts (4,6). Almost all cases with rosettes also had apparent acidophilic fibrillary intercytoplasmic background, which was the most useful diagnostic feature by light microscopic observation (7).

ON is commonly confused with undifferentiated carcinoma, lymphoma, melanoma or sarcoma. The advent



Figure 3. CT scan showing extensive nasal mass.

of immunohistochemistry has often provided greater accuracy in the pathological diagnosis of these neoplasms. Neuron specific enolase is always present and, in most cases, S-100 protein as well (5).

The diagnosis of ON by electron microscopy was mainly based on neuronal processes, neurosecretory granules, microtubules, neurofilaments, synaptic-like junctions and occasionally both microvilli and olfactory vesicles at the luminal borders of Flexner type rosettes (8). Moreover, the presence of catecholamines in ON provides a great help in diagnosis as does electron microscope examination (2).

Although other authors have noted a predominance of male patients, Fisher concluded that there was no sexual predilection for the tumor (1). The two major symptoms, epistaxis and nasal obstruction, occurred with equal frequency. On occasion they may appear as bleeding nasal polyps. The majority of these tumors arise in the upper nasal cavity, above the middle turbinate and extend into adjacent paranasal sinuses. These tumors can spread through the cribriform plate to involve intracranial contents at the skull base (8). Extracranial metastasis of ON is relatively uncommon. They are rarely found in cervical lymph nodes, lungs and bones; with 12.5% having intracranial extension. The 5- year survival rate of patients with ON without metastasis or intracranial extension is 50 % (5).

Kadish et al. (9) found the staging system to be a valuable predictor of survival where all stage A patients were disease free, while only 40 % of stage C patients remained so. Other studies have also found no relationship between clinical stage and survival.

Recent recommendations for therapy of olfactory neuroblastoma are for surgical extirpation followed by local radiation and/or chemotherapy, although some authors suggest that, using the staging method of Kadish et al. (9) stage A or B disease was equally well treated by surgery or radiation alone. Chemotherapy for more advanced disease may be effective in some cases if comparison to non-olfactory neuroblastoma is useful (3).

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