

OLFACTORY NEUROBLASTOMA: CLINICAL PRESENTATION AND MANAGEMENT

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ABSTRACT

Olfactory neuroblastoma (ONB) is a rare tumor. There is greater agreement for separating “neuroblastoma-like” forms from tumors arising in the same location and manifest morphological and immunohistochemical characteristics of neuroendocrine carcinoma.

The aim of present study is to describe the typical clinical, histological and immunohistochemical findings of ONB in 58-year old woman which help us to exclude non-Hodgkin's lymphoma and endocrine carcinoma.

Key words: olfactory neuroblastoma, neuroblastoma like forms, neuroendocrine carcinoma

Olfactory neuroblastoma (ONB) is a rare tumor first described by the French authors L. Berger et al. in 1924 /2/. It is a malignant neuroectodermal tumor assumed to originate from olfactory receptor cells high in the olfactory plate or in dystopic areas /1/. Most commonly the tumor is situated in the nasal cavity beyond the middle concha, and its extraolfactory localization is seldom /1/. The tumor growth is usually slow and over time infiltration of adjacent structures takes place in the direction of the paranasal sinuses, oro-nasopharynx, the base of skull and even the cerebral frontal lobes. Metastatic spread is locally to cervical lymph nodes, distally to lungs and bones and through leptomeninges into the brain /3/.

ONB occurs in a broad age range from 2 to 90 years without any gender predilection. The age of onset is to some extent correlated with the morphological pattern of ONB: tumor showing the more classical neuroblastic Homer-Wright rosettes tend to arise at an earlier age /4/. Most of the tumors are immunoreactive for neuron-specific enolase (84 – 100%), synaptophysin (64 – 100%) and 1/3 of the cases are positive for CK MNF /4/.

CASE REPORT

A 58-year old woman was admitted to the Department of Ear, Nose and Throat Diseases with symptoms of difficult nose breathing, recurrent bleeding from the right nostril, strong headache with frontal localization, smell dis-

turbances, slurry and decrease vision with right eye, diplopia. Two years before she had been diagnosed with “Pansinusitis polyposa” in another hospital. However, her complaints persisted and became more intensive, especially for the often recurrent bleeding from the nostrils. The patient developed anemic syndrome (normochromic, with normocytosis). She suffered from arterial hypertension, diabetes mellitus type II with obesity, diabetic polyneuropathy, chronic bronchitis.

On examination: BP 160/100, 130/80. Pulse 88. Auscultatory phenomenon for cor hypertonicum. Lab. tests: Hb 92, (86), 100 after hemotransfusion. Ht - 0.26, Er - 3.2, gluc. - 8.6, Chol. - 5.6, 3-glic. - 2.2.

Neurological status: Ipsilateral (right-sided) anosmia, primary optic subatrophy, contralateral praeretinal haemorrhagiae. Lesion of the left VI cran. nerve with diplopia.

CT description: tumor in the nasal cavity, in the maxillar, ethmoidal and sphenoidal sinuses with destruction of the sella turcica.

A bilateral ethmoidectomy was performed in the Department of Ear, Nose and Throat Diseases. The surgeon found out heavy bleeding tumor-like formations with smooth surface and soft texture. No enlargement of cervical and submandibular lymph nodes was established. The operative and postoperative periods were abnormal – with recurrent heavy bleeding from the nose and in the subarachnoid space 48 hours after the operation. The patient developed sopor, meningeal symptoms, left sided central hemiplegia, bilateral Babinsky sign, left Openheim sign, and skew deviation of the eyes on the right. After treatment in the Intensive care dept. the listed symptoms disappeared, but hemiplegia persisted. CT contrast examination before discharge showed: tumor persisting in the nasal cavity, paranasal sinuses, epipharynx and cellar region. The subarachnoid haemorrhage was fully resorpted.

The histological examination of the material from the ethmoidal sinus revealed a polypous formation with oedema and scanty inflammatory infiltration. Only in a small portion there was an olfactory neurogenic tumor. The light-microscopic study of the histological slides stained with

hematoxylin-eosin and Van Gieson, demonstrated the following histological features: cellular neoplasm, composed of small uniform cells with scant fibrillar cytoplasm and dark round nuclei. Tumor cells were arranged in lobules, containing scattered rosettes of Homer-Wright type. The tumor stroma was scanty and presented mainly of connective tissue with numerous capillaries. The neoplastic cells were immunoreactive for both: neuron-specific enolase (NSE) and cytokeratin.

DISCUSSION

The olfactory neuroblastoma is of morphological interest not only as a rare tumor, but also because it's diverse histological appearance, encompassing a spectrum of morphological subtypes. More recently, there has been

greater acceptance for separating "neuroblastoma-like" forms from tumors that arise in the same location and manifest morphological and immunohistochemical characteristics of neuroendocrine carcinoma /3/. In the case reported here, we made the diagnosis on the base of typical histological features and immunohistochemical findings, which help us to exclude non-Hodgkin's lymphoma and endocrine carcinoma.

We regard that recurrent nasal bleedings leading to anaemia in combination with visual disturbances must be very suggestive for neuroblastoma. Even routine polypectomy requires a histological examination, which can be combined with immunohistochemistry for a more precise diagnosis. ONB in advanced stages is unable for radical operative treatment.

REFERENCES

1. Головин Д. И. Ошибки и трудности гистологической диагностики опухолей. Москва, Медицина, 1982: 120. (in Russian)
2. Райчев Р. Биопсия - морфологична интерпретация. София, Мед. и физк., 1984:66. (in Bulgarian)
3. Kleihues P., Cvenec W. K. World health organisation classification of tumours. Pathology and genetics of tumours of the nervous system. IARC Press, Lyon, 2000: 150 – 152.
4. Mills S. E., Fechner R. E. The nose, paranasal sinuses and nasopharynx. In: Sternberg S. S. editor. Diagnostic surgical pathology, 2nd ed., New York, Raven press, 1994: 866 – 869.

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