Olfactory Neuroblastoma - Case Report

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Introduction

The olfactory neuroblastoma oresthesioneuroblastoma, is a tumor pathologically related to but clinically distinct from the childhood neuroblastomas originating from the adrenal medulla and sympathetic chain[1] - This tumor pnmarily affects patients between the second and fourth decades of life and most frequently originates in nasal cavity from the olfactory neuroepithelium[1]. Rare cases have been described to be arising in the nasophaiynx, maxillary sinus and ethmoid sinus[2] - The clinical course is characterized by local aggressiveness and less commonly distant metastasis. The tumor is relatively more radio-responsive than childhood neuroblastoma[1]. Identification of olfactory neuroblastoma from undifferentiated carcinoma and other intranasal malignancies is done by using the histological criteria[3] and demonstration of specific markers such as neurofilament and neuron-specific enclose (NSE) by immunohistochemical methods[4]. Electron microscopy may be used but has been infrequently employed[1].

Case Report

A 30 years old woman was admitted to the Jinnah Postgraduate Medical Centre (JPMC), Karachi because of recurrent epistaxis, pain, protuberance of left eye and nasal obstmction. Physical examination revealed a large fleshy highly vascular mass filling the left nasal cavity.

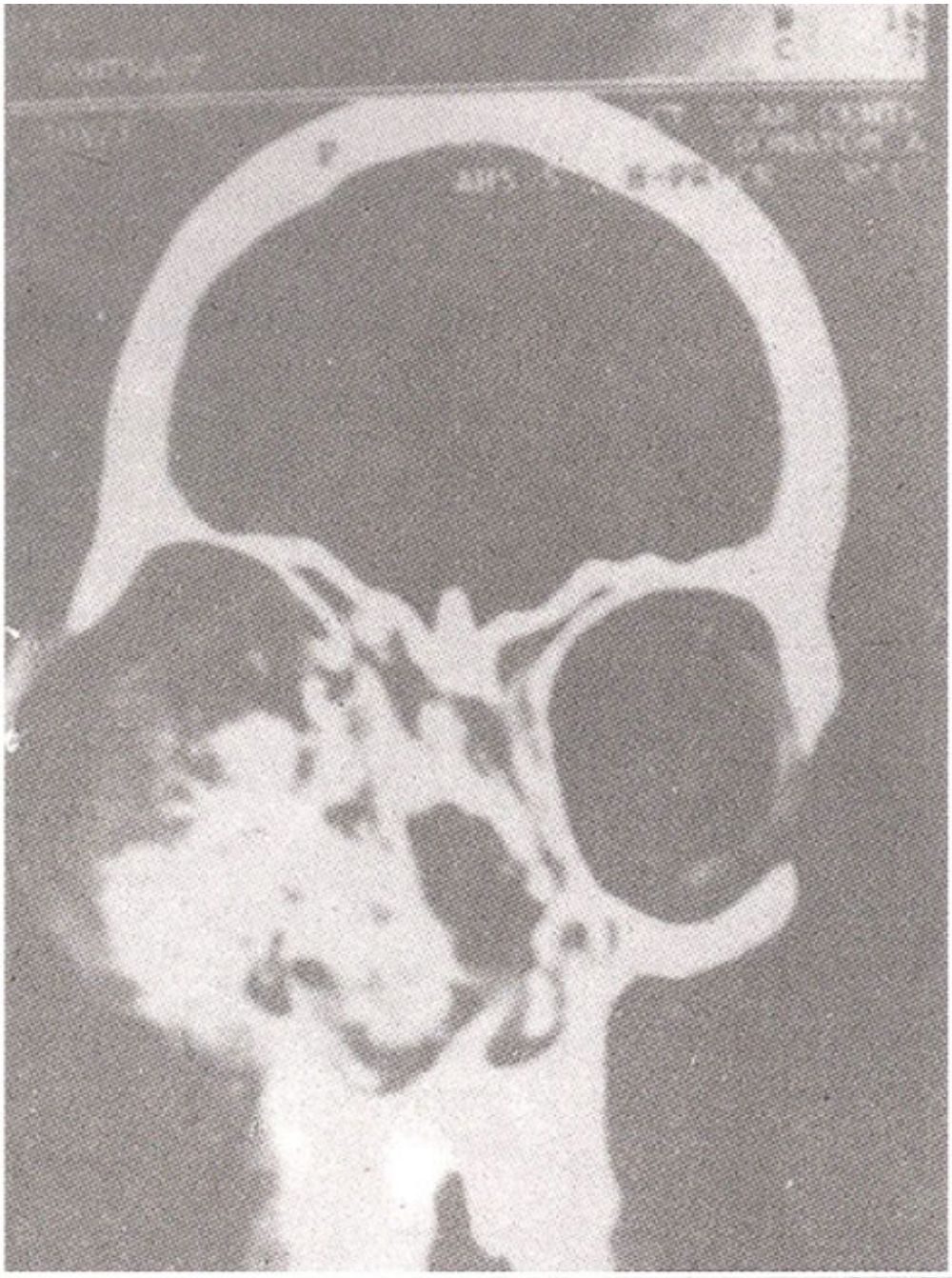

Figure 1. CT scanning revealing a large expansile destructing lesion involving the alveolar ridge of left maxillary bone, left maxillary sinus and nasal area.

CT scan (Figure 1) showedlarge expansile destructive lesioninvolving the alveolar ridge of left maxillary bone, left maxillary sinus and nasal area. The mass had also involved the ethmoid and sphenoid sinuses. No intracranial extension was seen. Punch biopsy was done and sent to the Department of Pathology where it was processed as for routine paraffin embedding. Sections were cut and stained by Haematoxylin and Eosin. A histological diagnosis of olfactory neuroblastoma was made. Sections revealed a tumour composed of groups of small monotonous cells with vesicular nuclei, rare mitosis and indistinct cell margins in an abundant finely fibrillary background (Figure 2).

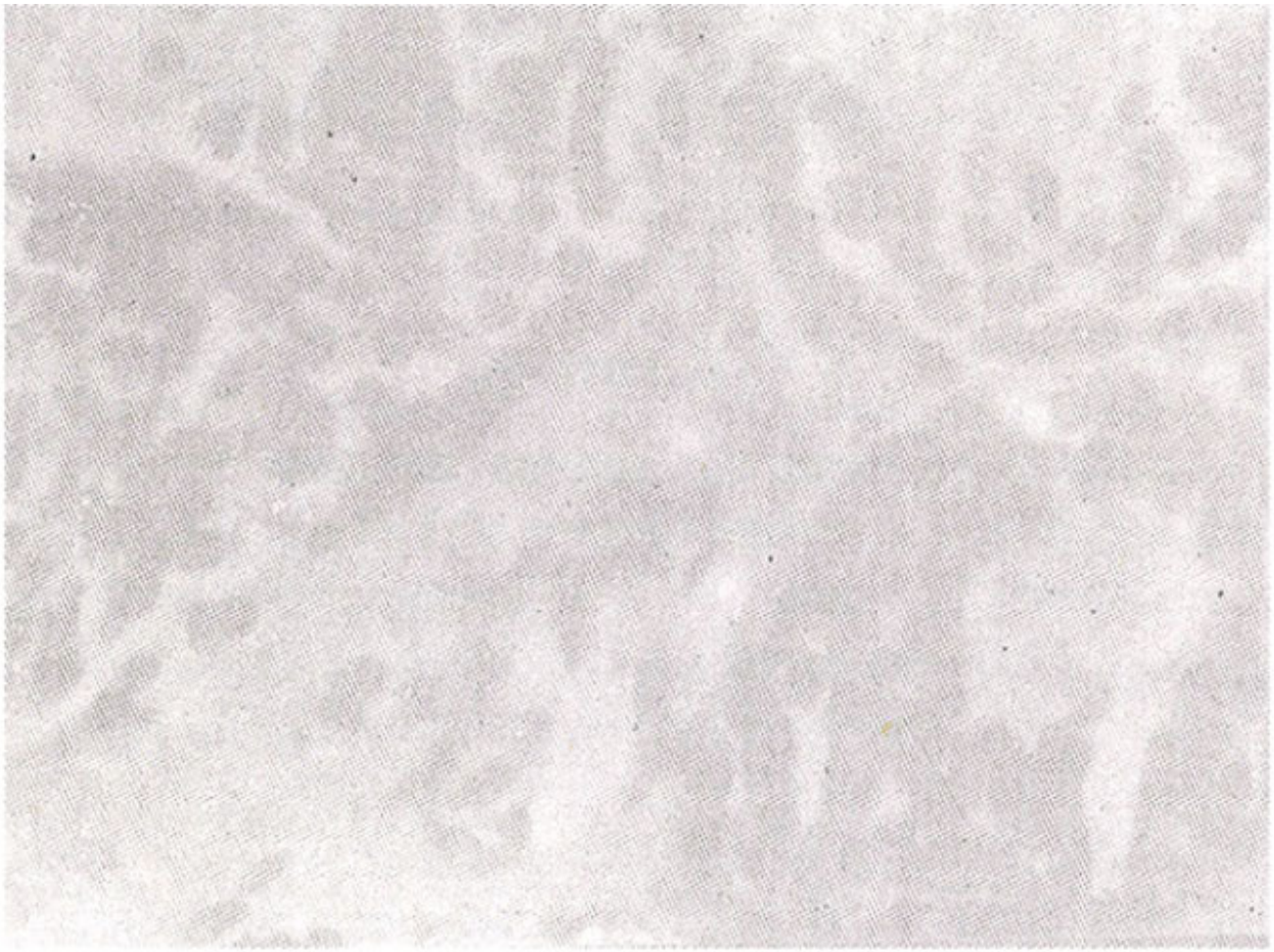

Figure 2. Histopathological section of a tumor consisting of a group of small, monotonous cells with vesicular nuclei, rare mitosis and indistinct cell margins growing in an abundant finely fibrillary background.

The monoclonal antibodies against the neurofilament stained scattered neoplastic cells within the islands of tumour and did not stain any of the non-neoplastic elements in the tumor such as fibroblast or blood vassels (Figure 3).

Figure 3. Shows tumour cells heavily to moderately stained by monoclonal antibodies against the neurofilament (arrows).

However, attemp to stain the tumor cells with monoclonal antibodies against neuron-specificenclose (NSE) was unsuccessful.

Discussion

The numberof olfactory neuroblastomas seen at any one institution is few and therefore, conclusions reached by various authors are not always congruous[5]. Olfactory neuroblastoma occurring primarily in the maxillary sinus are rare[1], but are still consistent with an origin from the olfactory epithelium, since the normal distribution of the olfactory epithelium may extend to the level of the middle turbinates. Also irregular demarcations between olfactory and respiratory epithelium are known to occur with age[1] and the ectopic localization of olfactory epithelium in the maxillary sinus is not inconceivable. On light microscopy several patterns have been described[6,7]. In this case the most easily recognizable and common appearance was seen, that is of a cellular tumor composed of uniform small cells with round nuclei, scanty cytoplasm, indistinct nuclear membranes and a prominent fibrillary background with areas of calcification. In addition, the presence of neurofilarnent protein was demonstrated in these tumor cells. This data provides strongest evidence of neuronal originof the tumor since neurofilamentproteins have been described only in neurons and neuronal cell lines[4]. The failure to demonstrate the neuron- specific enclose in the tumor cells may be due to the malignant transformation of these tumor cells which may be associated with an aberrant neuron-specific enclose synthesis.

Review of literature shows that unlike neuroblastomas arising in other sites, olfactory neuroblastomas occur over a wide age range[1]. Their biologic behaviour is unpredictable, some being extremely aggressive and others relatively indolent[5]. Although some of these tumours may be radiosensitive, probably none are radiocurable. Maximum therapeutic benefit seems best achieved with total gross removal followed by a full tumoricidal dose of radiation[3].

References

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