

BDIAP November Meeting 2010, Slide Seminar.

Case 8. Dr. Roderick HW Simpson.

Clinical History.

Male 52.

2010 May Epistaxis. MRI scan shows a large mass on the right involving most of the nasal cavity.

2010 June Biopsy.

Microscopic findings.

The specimen comprises oedematous sinonasal mucosa containing islands of tumour cells, set in a vascular stroma. The tumour cells consist of round to oval, regular nuclei with scanty pale cytoplasm. Mitotic figures are scanty. Rare possible rosettes or pseudo-rosettes are noted. There are small amounts of neurofibrillary matrix. No necrosis is seen.

Immunohistochemistry.

Synaptophysin+++ . Chromogranin A+++ , CD56+++ , Vimentin +++ , NSE +++ .

S-100 (only surrounding sustentacular cells).

cytokeratins (MNF116, CAM) focally + in the tumour, as well as nasal glands.

Desmin, p63, GFAP, neurofilament protein are all negative.

Diagnosis.

Nose-olfactory neuroblastoma (Hyams grade 1-2).

Clinical course.

2010 Aug: MRI shows a large mass in the upper part of the right nasal cavity extending into the medial aspect of the maxillary antrum and up into the ethmoid air cells. There is no breach of the cribriform plate.

2010 Sep: R lateral rhinotomy. Tumour found to involve cribriform plate.

2010 Oct: Radiotherapy planned.

Discussion.

Olfactory neuroblastoma is a malignant neuroectodermal tumour thought to originate from the olfactory membrane of the sinonasal tract. Patients range in age from 2-90 years, with two peaks in 2nd and 6th decades. The usual site is the upper nasal cavity in the region of the cribriform plate.

Typically the tumours are localised to the submucosa, growing in circumscribed lobules or nests separated by a richly vascularised fibrous stroma. The neoplastic cells have uniform small round nuclei with scant cytoplasm, dispersed chromatin and inconspicuous nucleoli. Nuclear pleomorphism, mitotic activity and necrosis are usually seen only in high grade tumours. The cells do not have distinct borders, and they are surrounded by a neurofibrillary matrix.

The majority of cells express synaptophysin and Chromogranin A, with S-100 reacting just with the surrounding sustentacular cells. About 10% of cases show a few cells staining for cytokeratin.

The Hyams' grading system recognises four grades, based principally on architecture (mainly degree of lobularity), cytological pleomorphism, neurofibrillary matrix, rosette formation, necrosis and mitotic activity.

The differential diagnosis includes other poorly differentiated sinonasal tumours, such as lymphoma, melanoma, SNUC, small cell neuroendocrine carcinoma and PNET. These can largely be distinguished by use of a panel of immunohistochemical markers.

The prognosis olfactory neuroblastoma largely depends on clinical stage, in particular the local extent. Therefore, tumours confined to the nasal cavity have a 75-91% five year survival, whereas the comparable figures for those extending beyond the nose and sinuses is 41-47%. Other factors said to be prognostically important include histological grade and the proliferation index.

References.

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3. Prasad ML, Perez-Ordoñez B. Non-squamous lesions of the nasal cavity, paranasal sinuses and nasopharynx. Chapter 3 in Gnepp DR, ed. *Diagnostic Surgical Pathology of the Head and Neck*, 2nd edn. WB Saunders, Philadelphia 2009: 148-154.
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