Olfactory Neuroblastoma and Olfactory Ventricle

A Case Report

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SUMMARY – *A case of dumbbell nasal and intracerebral olfactory neuroblastoma (ONB) related to possible seeding through an “olfactory ventricle” in the olfactory bulb and nerve is reported. This anatomic variant was recognized at operation for the first time: it consists of a hollow cavity within the olfactory bulb and nerve which may be connected to the subventricular zone of the frontal horn of the lateral ventricle. Better scrutiny of this structure is necessary in view of its possible interference with the course and diffusion of ONBs.*

Introduction

Olfactory neuroblastoma (ONB) is a rare malignant upper nasal cavity tumour often involving the anterior cranial fossa. It arises from a specialized epithelium in the nasal mucosa and in particular from the spherical basal cells which constitute a stem cell compartment². This olfactory neuroepithelium is a unique neurosensory organ, because olfactory neurons are continuously replaced throughout adult life^{2,3}. The ONB spreads to the anterior skull base through the lamina cribrosa parasitizing the olfactory bulb (OB) and tract. The unusual diffusion of an ONB to the brain is generally due to haematogenous or leptomeningeal dissemination and corresponds to the T4 stage of the ONB staging system². We describe a case of a dumbbell nasal and intracerebral ONB related to possible tumoral seeding through an “olfactory ventricle” which consists of a hollow cavity within the OB and nerve which may be connected through a tubular extension to the frontal horn of the lateral ventricles¹.

Case Report

While on holiday abroad, a 45-year-old Italian woman complained of headache, vomiting and decreased level of consciousness and was

immediately taken to the nearest hospital. CT scan and MRI on admission revealed a huge haemorrhagic and partially calcified mass in the left frontal lobe (Figure 1) which was immediately excised through a left frontal craniotomy. The pathologic diagnosis was problematic and a presumptive diagnosis of gliosarcoma or primitive neuroectodermal tumour was made. The post-operative course was uneventful and the patient returned to Italy after 15 days. Awaiting the definitive diagnosis, no postoperative adjuvant therapy was administered. After two months she complained again of headache and slurred speech: MRI demonstrated a dumbbell lesion constituting a huge recurrence of the left frontal lesion and by a mass occupying the naso-ethmoidal region apparently in continuity through the lamina cribrosa (Figure 2 A,B). A craniofacial approach was planned and both tumours were completely resected. At operation, the two lesions were connected by a strand constituted by a thickened left olfactory bulb. A hollow right OB was also noted (Figure 3). The pathological diagnosis was ONB both in the nose and brain (Figure 4). Looking better at pre-operative MRI, we recognized a hollow right OB and nerve (Figure 2C), resembling a rare anatomic variant called “olfactory ventricle”⁸. Postoperative radio-chemotherapy was administered (60 Gy in 30 fractions and platinum/etoposide phosphate). In the following months

Figure 1 Post-contrast axial T1-weighted MRI performed on admission to a foreign hospital, showing a left frontal mass presenting with cystic and solid areas. The tumour was promptly excised.

her condition remained satisfactory. Unfortunately, eight months later MRI demonstrated diffusion of the disease to the brain and spinal cord leading to a rapid clinical deterioration.

Discussion

The lateral wall of the human lateral ventricles, the so-called subventricular zone (SVZ) contains a ribbon of astrocytes with neural stem cell properties⁶. In rodents and some mammals these newly born subventricular zone progenitors reach the OB through a pathway called the rostral migratory stream (RMS): until recently, its demonstration in adult human brain has been elusive⁴. Curtis et al. recently identified a previously undefined structure in the human forebrain which was named the ventriculo-olfactory neurogenic system (VONS) consisting of the SVZ, the rostral migratory stream and the olfactory tract and bulb. They also demonstrated in the olfactory bulb and tract a hollow cerebrospinal fluid-filled cavity they termed "olfactory ventricle" connected to the lateral ventricle through a tubular extension within

the RMS^{1,3}. These findings were confirmed by MRI showing, as in our case, a tube-like structure measuring about 7mm in length and 1.5 mm in width within the OB. The human RMS found by Curtis et al. also contained progenitor neural cells with migratory characteristics which may become mature neurons in the OB¹.

The olfactory ventricle exists early in human development and closes during foetal growth⁷: in adults, it probably represents a remnant of the original embryonal structure which may be associated with a defective maturation of the olfactory pathways. Although the existence of the olfactory ventricle has been questioned and its incidence is still debated^{1,7}, our case suggests that it may be related to a brain tumour originating along the olfactory pathways. The aggressive course of the ONB and the presentation as a haemorrhagic intracerebral mass lesion is unusual. This tumour generally invades the anterior cranial base through the lamina cribrosa: its dumbbell appearance with a distinct intracerebral haemorrhagic component is rare⁵. In our case the intracerebral lesion may have resulted from seeding of the original nasal tumour through the olfactory ventricle and its

Figure 2 Post-contrast sagittal (A) and coronal (B) SE T1-weighted MRI two months later show a heterogeneous dumbbell enhancing tumour presenting with cystic and solid areas. The lesion occupies the upper nasal cavity, erodes the cribriform plate, expands into the anterior cranial fossa and involves the frontal lobe through a basal infiltration corresponding to the olfactory nerve. The intact hollow right olfactory nerve is visible (arrow). C) Axial T1-weighted MRI better shows the hollow right olfactory bulb and nerve (arrow) containing a hypointense thin cavity found at operation. The left olfactory nerve is infiltrated by the tumour and is not fully recognizable.

Figure 3 Intraoperative picture showing a sectioned hollow right olfactory bulb (arrow) representing the so-called "olfactory ventricle".

Figure 4 A) On H&E (100× magnification) the tumour is constituted by round uniform cells with inconspicuous nucleoli, surrounded by delicate capillaries. Cells are embedded in a delicate fibrillary background, composed of neuropils, and (B) show some areas of rosetting. (C) The nests of neoplastic cells were immunopositive with anti-synaptophysin antiserum (100× magnification).

frontal extension along the olfactory pathways. Alternatively, the independent growth of two distinct lesions may occur owing to an “ectopic” intracerebral origin resulting from defective migration and aberrant growth of neuroblasts along an abortive RMS in the frontal lobe. The olfactory ventricle and its connection to the lat-

eral ventricle via a continuous open ventricle may constitute an unusual route of contamination and vulnerability of the cerebrospinal fluid and central nervous system through the nose: better scrutiny of this structure is necessary in view of its possible interference with the course and diffusion of ONBs.

References

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