# Olfactory bulb hamartoma

## Case series

Brain MRIs of 110 tuberous sclerosis complex (TSC) patients (mean age 11.5 years; age range 0.5-38 years; 52 female; 26 TSC1, 68 TSC2, 8 without mutation identified in TSC1 or TSC2, 8 not tested) were retrospectively evaluated. Signal and morphological abnormalities consistent with olfactory bulb hypo/aplasia or with olfactory bulb hamartomas were recorded. Cortical tuber number was visually assessed and a neurological severity score was obtained. Patients with and without rhinencephalon abnormalities were compared using appropriate parametric and non-parametric tests.

Eight of 110 (7.2%) TSC patients presented rhinencephalon MRI changes encompassing olfactory bulb bilateral aplasia (2/110), bilateral hypoplasia (2/110), unilateral hypoplasia (1/110), unilateral hamartoma (2/110), and bilateral hamartomas (1/110); olfactory bulb hypo/aplasia always displayed ipsilateral olfactory sulcus hypoplasia, while no TSC patient harboring rhinencephalon hamartomas had concomitant forebrain sulcation abnormalities. None of the patients showed overt olfactory deficits or hypogonadism, though young age and poor compliance hampered a proper evaluation in most cases. TSC patients with rhinencephalon changes had more cortical tubers ($47 \pm 29.1$ vs $26.2 \pm 19.6$; $p = 0.006$) but did not differ for clinical severity ($p = 0.45$) compared to the other patients of the sample.

Olfactory bulb and/or forebrain changes are not rare among TSC subjects. Future studies investigating clinical consequences in older subjects (anosmia, gonadic development etc.) will define whether rhinencephalon changes are simply an imaging feature among the constellation of TSC-related brain changes or a feature to be searched for possible implications in the management of TSC subjects [1)].

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Gross or microscopic glial hamartomas were found in the anterior olfactory lobe and olfactory germinal layer of three babies, two of them newborns, with tuberous sclerosis. In two cases microscopic hamartomas were seen in the anterior olfactory lobe, and in one of them there was a prominent nodular tumor of the olfactory tract and trigone. In addition, in both of these cases there were bilateral germinal layer tumors between striatum and septum, at the junction of the obliterated olfactory recess and the frontal horn of the lateral ventricle. Only microscopic hamartomas were present in the olfactory germinal layer of the third case. Typical subependymal germinal layer tumors were also present elsewhere in all cases; however, cortical tubers were recognized in only two of them. In all three patients, the clinical presentation and death were due to cardiac rhabdomyomas. The findings suggest that olfactory hamartomas might be relatively common in tuberous sclerosis. Involvement of olfactory structures is not surprising because the lesions seem to originate in the germinal layer, a region of the brain which is prominently involved in the disease [2)].

## Case reports

The objective of a case report was to demonstrate that a benign tumour arising solely from the olfactory bulb can act as a primary epileptic focus and to illustrate the difficulty in making a preoperative diagnosis of pathological lesions in this area. The case of a 29-year-old male with

intractable epilepsy is presented. Radiological imaging demonstrated a conspicuous, calcified lesion in the mesial inferior frontal gyrus. Electroclinical findings confirmed this to be the epileptogenic zone. Surgery and subsequent histology surprisingly revealed this focal lesion to be a benign hamartoma arising solely from the olfactory bulb. Resection resulted in seizure resolution. Tumours of the olfactory apparatus are extremely rare, but can present with epilepsy [3].

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2)
de León GA, Zaeri N, Foley CM. Olfactory hamartomas in tuberous sclerosis. J Neurol Sci. 1988 Nov;87(2-3):187-94. PubMed PMID: 3210031.
3)
McEvoy AW, Bartolucci M, Revesz T, Harkness W. Intractable epilepsy and olfactory bulb hamartoma. A case report. Stereotact Funct Neurosurg. 2002;79(2):88-93. PubMed PMID: 12743430.