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Tongue-base hamartoma in tuberous sclerosis

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Abstract
This paper describes the case of a 41-year-old female with tuberous sclerosis who persented with a large tongue-base hamartoma. The surgical management of the patient was complicated by the presence of a large thyroid goitre. Awake fibre-optic intubation, thyroidectomy then tracheostomy were necessary before the tongue-base hamartoma could be safely resected. To the best of our knowledge, this is the first reported case of a tongue-base hamartoma in a patient with tuberous sclerosis.

Key words: Tuberous Sclerosis; Tongue Neoplasms; Hamartoma

Case report

A 41-year-old female presented with a one-year history of increasing stridor, voice change and symptoms of obstructive sleep apnoea. She also had a history of long-standing epilepsy and adenoma sebaceum. The diagnosis of tuberous sclerosis had been previously debated. In addition, 20 years previously, she had undergone thyroid surgery for thyrotoxicosis. Toxic symptoms recurred 10 years later and she was treated with radioactive iodine.

Clinical examination revealed biphasic stridor, hot potato speech, a very large goitre and also a large, fronded mass at the tongue base which obscured the larynx. Computed tomography (CT) scan showed a multinodular goitre largely involving the left thyroid lobe (Figure 1), distortion of the larynx above the vocal folds and a large mass arising from the tongue base (Figure 2).

For surgery, the airway was secured by nasal fibre-optic awake intubation in the sitting position. Even using this technique, intubation was very difficult. The old thyroidectomy scar was re-opened and the trachea was exposed by removal of the grossly enlarged isthmus and

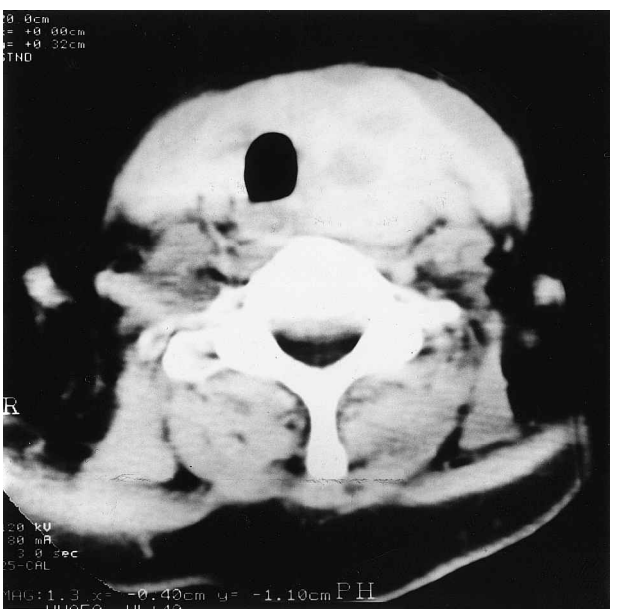

Fig. 1
Axial CT scan showing the goitre.

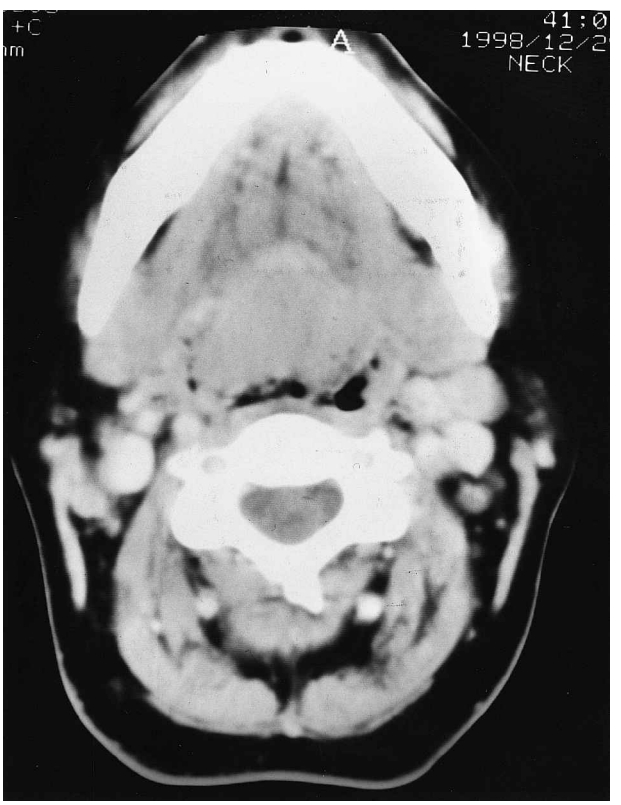

Fig. 2
Axial CT scan showing the mass arising from the tongue base.

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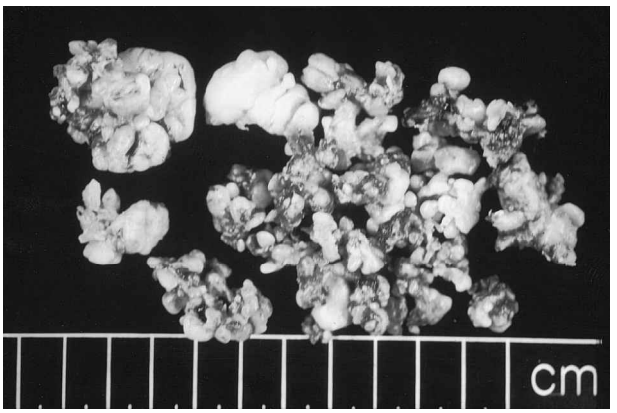

FIG. 3
Tissue removed from the tongue base.

left thyroid lobe. A tracheostomy could then be performed. Pharyngoscopy revealed a massive proliferative lesion of the tongue base, that was endoscopically debulked (Figure 3).

Two further tongue-base debulking procedures were subsequently performed and the patient has since had the tracheostomy removed. She has no residual stridor nor obstructive sleep apnoea and her speech is much improved.

Histology of the thyroid gland tissue confirmed a multinodular goitre. The tissue removed from the tongue base at the first operation weighed 30 grams and measured $7 \times 7 \times 2$ cm. The histology of this tongue-base lesion showed reactive tonsillar tissue; a hamartoma of the tongue base.

Discussion

Tuberous sclerosis complex (TSC) is a rare autosomal dominant neurocutaneous disorder with a high spontaneous mutation rate.[1] TSC results in hamartomas affecting many organ systems. Organs most commonly affected are the brain, skin, heart and kidney.

TSC was previously thought to present as the classic triad of epilepsy, adenoma sebaceum and mental retardation. Diagnostic criteria have recently been revised in the light of new clinical and genetic information.[1,2] No single sign is present in all affected patients, nor is any one clinical or radiological sign absolutely specific for TSC. Clinical features are now categorised as major and minor features depending on their specificity for TSC. Patients are categorised as having definite, probable or possible TSC depending on which clinical signs they have.[2]

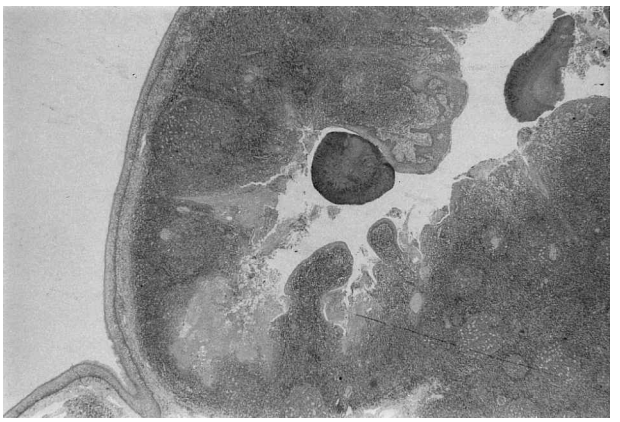

FIG. 4
Histological section of tissue removed from the tongue base; characteristic tonsillar crypts containing Actinomyces are observed (H&E; ×45.7).

This patient fits the revised diagnostic criteria for definite TSC. Non-renal hamartomas (e.g. tongue-base hamartoma) are classified as a minor feature of TSC. To the best of our knowledge this is the first report of a tongue-base hamartoma in a patient with TSC.

This patient posed a management problem. Her airway was severely compromised by the tongue-base mass which obscured the larynx. This made an awake fibre-optic nasal intubation by a senior anaethestist a necessity. Tracheostomy was necessary before the tongue base could be safely resected and removal of the large goitre was needed to expose the trachea. Tongue-base debulking provided the histological diagnosis and relieved the patient's symptoms.

This patient also has a long history of florid thyroid disease. A literature search provided no evidence to link thyroid disease with TSC.

References

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Miss H. Wallace takes responsibility for the integrity of the content of the paper.
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