The Journal of Laryngology and Otology
December 1998, Vol. **112**, pp. 1189–1191

An unusual cause of obstructive sleep apnoea presenting during pregnancy

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Abstract
We describe a case of lingual thyroid (LT) with primary hypothyroidism, presenting during pregnancy and continuing beyond it with oropharyngeal obstructive symptoms and sleep apnoea syndrome (SAS) of mixed type. Although SAS of a combined obstructive and central type should not be too surprising in a case of LT with hypothyroidism, we were unable to find such a documentation previously. Only four weeks of L-thyroxin treatment resulted in a dramatic improvement in dysphagia, disturbed phonation, haemoptysis, arterial desaturation, sleep apnoea and overall sleep efficiency, in conjunction with a regression in the size of the lingual mass. This case highlights the vagaries confronted in the management of such a case and focuses on efforts towards accurate diagnosis and treatment.

Key words: Lingual thyroid; Sleep apnoea syndrome; Pregnancy; Hypothyroidism

Case report

A 22-year-old female was referred to our hospital for embolization of an oropharyngeal mass. A biopsy of the mass performed at the referring hospital revealed a diagnosis of a cavernous haemangioma. At the time of presentation to our clinic, the patient and her husband gave a history of progressively worsening snoring and obstructed breathing episodes occuring several times at night. She also complained of difficulty in swallowing and a change in her voice starting in the first trimester of her first pregnancy, which had terminated successfully two months ago. There was also a history of blood spitting, breathlessness on exertion and a foreign body sensation in the throat. The patient had a hardly comprehensible, slurred and slow (cu-de-sac, or hot potato) speech. Examination revealed a 3 × 3 × 1.5 cm fleshy mass protruding from the posterior third of the tongue extending into the valeculla and pushing the epiglottis posteriorly. Remainder of the examination did not show abnormal positive findings.

A magnetic resonance imaging (MRI) study with gadolinium confirmed the presence and the location of the mass. There was a large (6 × 4 cm) lobulated solid mass at the base of the tongue compromising the upper airway (Figure 1) with confirmation of the absence of a thyroid gland in the neck. The signal intensity of this mass was higher than that of the tongue on both T1 and T2 scans, and there was a further increase of the intensity after administration of gadolinium. The mass was best visualized on the T2-weighted images. A whole body radioactive iodine ^{123}I scan was carried out after establishing a negative pregnancy test. This showed a focal concentration of intense radioactivity at the base of the tongue (Figure 2) with a 24-hour ^{123}I uptake value of eight per cent. There was no demonstrable uptake of ^{123}I elsewhere in the body. Hormonal evaluation revealed evidence of primary hypothyroidism as the serum-free T4 concentration was 8.7 pmol/L (Reference range 10–19), T3 1.8 nmol/L

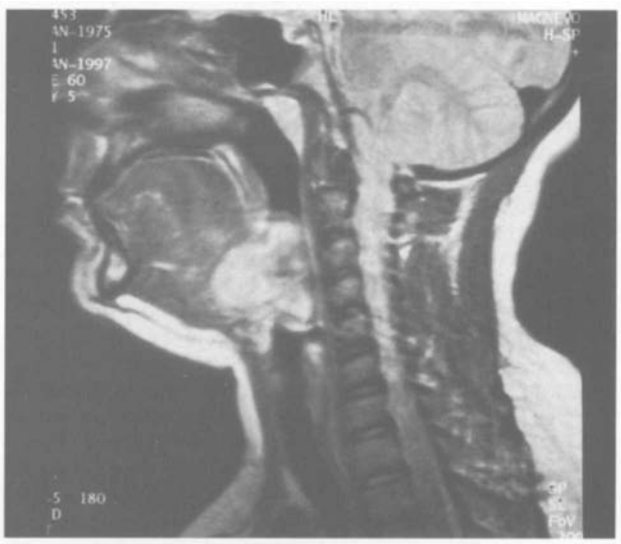

FIG. 1

MRI T2-weighted fast spin-echo sagittal scan shows a 3.5 × 4.2 cm high intensity signal mass in the posterior one third of the tongue protruding into the vallecula and oropharynx. The epiglottis is displaced posteriorly by the mass which is heterogeneous, sharply defined and obstructing the upper airway.

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Accepted for publication: 12 September 1998.

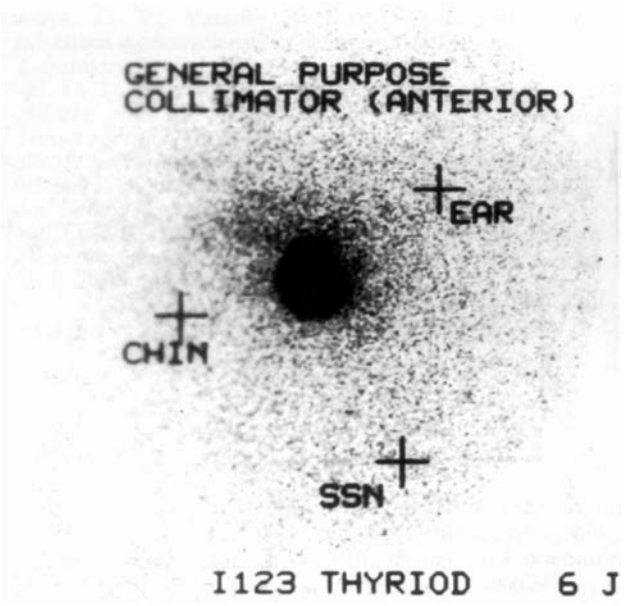

FIG. 2

^{123}I scan of the neck and oral cavity region showing a focal concentration of intense radioactivity localized to the posterior tongue. There is no uptake present in the neck where normally the thyroid gland is situated.

FIG. 3

Four months after thyroxin therapy gadolinium-enhanced T1-weighted spin-echo sagittal MRI scan shows a marked interval decrease in the size of the lingual thyroid. It now measures 1.9 × 2.1 cm, significantly clearing the airway.

(Reference range 1.0–2.5) and TSH 26.7 mU/L (Reference range 0.35–5.5). The antiperoxidase and anti-thyroglobulin antibodies were undetectable.

To evaluate the nocturnal obstructive symptoms, initially an overnight pulse oximetry study was done during sleep using a transcutaneous finger probe. The study showed significant desaturation during sleep with the mean oxygen saturation being 81 per cent (range 47 to 92 per cent). During a four-hour sleep period, 82 episodes of desaturation occurred. This was followed-up with a full multi-channel polysomnography. This study showed 34 episodes of apnoea of 10 seconds or longer over a two-hour period and demonstrated a combined pattern of obstructive and central apnoea resulting in a sleep apnoea index of 128 (abnormal if >5).

A diagnosis of lingual thyroid with hypothyroidism was made and the patient was started on 100 microgram (mcg) of L-thyroxin. Review of the patient four weeks later showed that there was resolution of dysphagia, dysphonia, and a dramatic improvement in her sleep apnoea. This correlated with normalization of the thyroid profile: FT4 19 pmol/L and TSH 1.5 mU/L and a regression in the size of the mass lesion on a follow-up MRI. The patient was reviewed at four months after thyroxin treatment and the MRI was repeated. It showed a marked decrease in the size of the lingual thyroid (Figure 3). A follow-up ^{123}I study performed while the patient was on LT4 treatment revealed faint uptake at the base of the tongue. On a follow-up polysomnography the mean oxygen saturation improved to 98 per cent and the sleep apnoea index to 29. The patient has now been followed up for 18 months. There is a slight residual swelling at the tongue base, which is not causing any obstruction to the aero-digestive tract. She is presently on 100 mcg of thyroxin and is completely symptom-free.

Discussion

Lingual thyroid is a rare developmental anomaly characterized by the presence of thyroid tissue in the midline at the base of the tongue, between the circumvallate papillae and the epiglottis. The true incidence of lingual thyroid is not known. It is estimated to be around 1:100,000 (Neinas *et al.*, 1973). Interestingly, in an autopsy study the incidence was found to be 10 per cent (Saulk, 1979). By about the mid-1980's some 400 cases have been reported (Baldwin and Copeland, 1988).

Lingual thyroid often occurs without symptoms (Kansal *et al.*, 1987). It has two main modes of presentation based on patient's age at presentation (Williams *et al.*, 1996). One group is that of infants and young children who suffer from failure to thrive and mental retardation and the lingual thyroid is detected in them on routine examination. The second consists of those at peripubertal ages and the lingual thyroid presents in them with symptoms of oropharyngeal obstruction and/or dysphagia. Hypertrophy of the gland occurring in an aberrant location results from an increase in serum TSH levels as a result of enhanced metabolic stress during puberty. A similar situation may occur in other instances of metabolic demands, such as pregnancy (as in our patient), infection or trauma (Williams *et al.*, 1996). There is a marked female preponderance with a male to female ratio of 4:1 to 7:1. Between 75–100 per cent of patients with symptomatic lingual thyroid have no other functional thyroid tissue (Kansal *et al.*, 1987). About 70 per cent will have varying degrees of hypothyroidism (Batsakis *et al.*, 1996). Hypothyroidism is a well recognized cause of SAS (Rajagopal *et al.*, 1984).

The diagnostic criteria required to arrive at the accurate diagnosis of lingual thyroid simply include the identification of a lingual mass at the base of the tongue between the epiglottis and circumvallate papillae, with demonstration of uptake of radioactive iodine in the location following the administration of a tracer dose. Most lingual thyroid glands contain histologically normal tissue and histological confirmation of the diagnosis is not necessary unless iodine or CT scanning is negative or malignancy is suspected (Saulk, 1979). Unenhanced CT scanning of the oropharyngeal area is reported to show a diagnostic appearance (hyperdensity) due to the iodine content of the thyroid

https://doi.org/10.1017/S0022215100142811 Published online by Cambridge University Press

tissue. MRI represents a new modality for assessing gland size and position. Aside from avoiding radiation exposure it produces excellent and characteristic images similar to the normal tissue which is higher in signal intensity than surrounding tissues on both T1 and T2 weighted images of the ectopic gland.

A large lingual mass may cause life-threatening obstructive symptoms; indeed neonatal asphyxial death has occurred (Hickman, 1869). Complications which can occur in a lingual thyroid include haemorrhage, infection, aero-digestive obstructive symptoms and malignancy. There have been rare reports of malignancy and at least 23 cases have been described by 1996, both papillary and follicular types developing in a lingual thyroid (Diaz-Arias *et al.*, 1992).

The clinical management of lingual thyroid depends on its size, the presence and severity of symptoms and complicating factors such as ulceration, haemorrhage, or malignancy. For patients with hypothyroidism, life-long replacement treatment using L-thyroxin is the mainstay of treatment. Even for those who have not yet developed hypothyroidism or obstructive symptoms, life-long thyroxin treatment should be given to prevent the onset of hypothyroidism, enlargement of the gland and diminish the risk of malignancy. Surgical excision of a lingual thyroid with autotransplantation may be considered in a rare patient whose obstructive symptoms fail to respond to the hormonal treatment.

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