EDITORIAL

A Lingual Thyroid Associated With Congenital Profound Sensorineural Hearing Loss

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Abstract:

This is a case report of an extremely rare case of a lingual thyroid associated with profound bilateral congenital sensorineural hearing loss in a Sudanese girl. To our knowledge this association has not been previously reported.

الملخص:

هذا تقرير حالة نادرة الحدوث لغدة درقية لسانية مرتبطة مع صمم حسي عصبي ولادي وخيم ثنائي الجانب لدى فتاة سودانية. حسب علمنا هذا الإرتباط لم يسبق الإبلاغ عنه.

Keywords: lingual thyroid, sensorineural hearing loss, Pendred's syndrome.

Background:

Thyroid disease in association with congenital sensorineural hearing loss is extremely rare. The well established association is Pendred's syndrome which accounts for 7.5% of all cases of deafness. Pendred's syndrome is a genetic disorder leading to congenital bilateral sensorineural hearing loss and a normally situated goiter with occasional hypothyroidism [1, 2, 3]. The outer and middle ear is usually normal, but in some cases it is associated with inner ear structural anomalies particularly Mondini's deformity. On the other hand lingual thyroid is relatively rare and is estimated to occur in 1 in 300 cases of thyroid disease[4]. However it represents the most common location for functioning ectopic thyroid tissue. Lingual thyroid, which occurs most commonly in females, is associated with absence of normally located thyroid in 70% of cases.[5]

In this report we present an extremely rare case of a lingual thyroid associated with congenital profound sensorineural hearing loss in a Sudanese girl. In spite of meticulous search in on-line literature and clinical literature, we could not find a report of a similar case.

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Case Report:

S.E.A. is a 29 years old Sudanese girl who lives in a rural area in the Gezira state. She presented with a longstanding sensation of a foreign body in the throat and intermittent choking attacks. She is profoundly deaf since birth with poor speech development. The main informants were the parents. On examination, her general condition was good. Local examination of the ear, nose and throat confirmed profound bilateral symmetrical sensorineural hearing loss and a rounded reddish lingual mass at the posterior third of the tongue. The mass measured about 2.5cm in diameter with smooth surface and healthy oral mucosal lining. Tuning forks test confirmed bilateral symmetrical sensorineural hearing loss and otherwise there was no gross anatomical abnormality. Thyroid was not felt by neck examination and systemic examination revealed no abnormality. Complete blood count and urine analysis were within normal limits. Thyroid hormones (TSH, T3, T4) were within normal range. TC99M thyroid scan was reported as active thyroid tissue with regular outlines and uniform radio-isotope distribution at the base of the tongue with absence of the thyroid tissue in the neck (Figure 1).

Axial C-T scan of the neck with contrast showed 2.24×2.08 cm homogenously enhanced soft tissue mass at the posterior part of the tongue encroaching upon the hyoid bone and no definite enhanced thyroid tissue in the neck. High resolution C-T scan of the temporal bones showed normally developed inner ear structures.

Pure tone audiometry (P.T.A.) showed bilateral symmetrical profound sensorineural hearing loss (Figure 2).

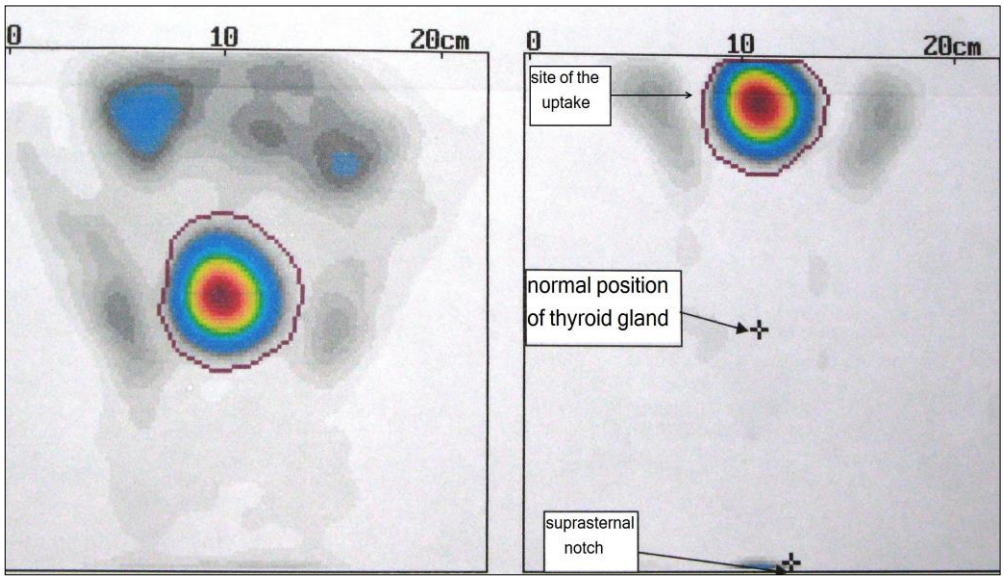

Figure (1): photograph of patient's thyroid TC99M radio-isotope scan.

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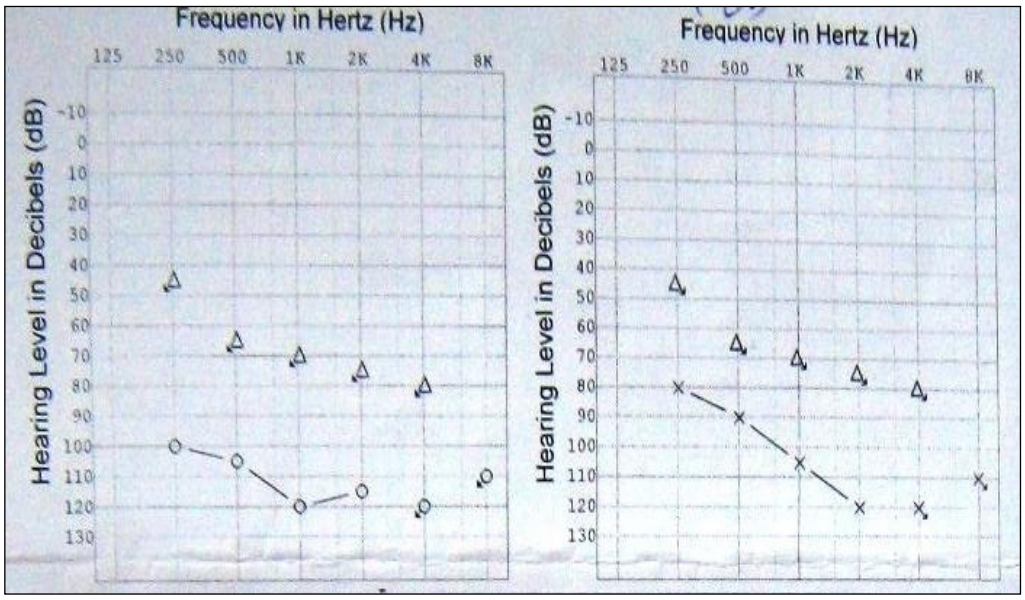

Figure (2): Pure tone audiogram (PTA) of the patient

Discussion:

The diagnosis of a lingual thyroid should be suspected when a mass is detected in the region of the foramen cecum of the tongue and is definitely established by radio-isotope scaning.(6)

The association of a goiter and sensorineural hearing loss is described as Pendred's syndrome and recent studies linked the disorder to mutations of the PDS gene which codes for the Pendren protein. This gene is located on the long arm of chromosome 7 (7q31). The disorder is inherited as autosomal recessive.(7)

In this case further genetic studies are needed to find out if there is any relation to Pendred's syndrome.

In some cases of Pendrid's syndrome the cochlear is dysplastic (Mondini dysplasia). However, in this study high resolution C-T scanning showed no abnormality of the cochlea.(8)

Hearing in Pendred's is usually abnormal, but findings are not particularly specific and an audiogram alone is not sufficient to diagnose Pendred's syndrome. Hearing loss is present since birth and language acquisition may be a significant problem.7

In conclusion we report this rare case of congenital sensorineural hearing loss associated with a lingual thyroid that may be similar to Pendred's syndrome. However, this case differs from Pendred's syndrome in that there is no goiter, but a lingual thyroid. The lingual thyroid is of a small size and the patient is euthyroid.

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