Case Report

Adult Cretinism with Lingual Thyroid presenting as Menorrhagia

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Abstract
Lingual thyroid is a rare disorder first described by Hickman in 1869. It has a prevalence of approximately 1 per 100,000 to 300,000 patients and female patients predominate in all. Only about 10% of patients with lingual thyroid can present as adult cretin. Here we present a case that despite the evidence of hypothyroidism, the diagnosis was delayed until the complaint of menorrhagia leading to severe symptomatic anemia led to its detection at the late age of 28 years.

Key words: Adult cretin, Lingual thyroid, Menorrhagia

Case Report
A 28 year female presented with complaints of menorrhagia for last 4 months. She had complaints of easy fatigability and excessive day time sleepiness for last 5 years. She never consulted any doctor for these complaints till four month back when she started with excessive menstrual bleeding approximately 35-40ml/day and it lasted for about 10 days. During next cycle patient again had excessive bleeding which lasted for about 8-9 days. She had achieved menarche at age of 19 years. She was born of full term normal delivery but her developmental milestones were delayed. She could speak and walk at around 6 years of age. She had not attended school and could only manage simple household work. There was no significant family history. On examination she was pale with periorbital puffiness, coarse hair with cold, dry and coarse thick skin. There was no thyromegaly and her otorhinolaryngological examination was normal. Secondary sexual characters were present (breast development at Tanner stage 3 and pubic hair at Tanner stage 4). She had a protrubent abdomen with epigastric hernia (Fig 1). She was clinically hypothyroid having delayed biceps and ankle jerks.
Her anthropometry revealed weight 30 kgs, height 128 cms (well below 3rd centile), arm span 120cms and head circumference 52 cms. The upper to lower segment ratio of 1.2, features compatible with cretinism (Fig 1).

Investigations revealed haemoglobin 6g/dl with normocytic normochromic peripheral film. Liver and kidney functions were normal. Fasting blood sugar and lipid profile was normal. Thyroid function tests revealed serum thyroxine 14ng/dl(60-200 ng/dl), serum thyroid stimulating hormone(TSH) 176µIU/ml (Normal 0.35-5.50µIU/ml) and anti-thyroid peroxidise antibody 146.30U/ml (Normal ≤60U/ml). Chest X-ray was showing mild cardiomegaly. ECG revealed sinus bradycardia @60bpm. Pure tone audiometry revealed normal bilateral hearing. Echocardiography showed mild pericardial effusion. Ultrasonography neck revealed hypoplastic thyroid tissue. Ultrasonography abdomen revealed cholelithasis, epigastric hernia and bulky uterus with thickened endometrium.

A technetium pertechnate thyroid scintiscan showed no significant uptake in the neck but showed a focus of tracer uptake at the root of tongue (Fig 2). In view of severity and apparent long duration of hypothyroidism together with the low body weight patient was started on low initial dose of thyroxine 12.5µg/d. This dose was gradually increased to current dose of 100µg/d.

Discussion
Lingual thyroid is a rare disorder first described by Hickman in 1869. It has a prevalence of approximately 1 per 100,000 to 300,000 patients and 1 in 4000 cases of thyroid disease. Female patients predominate in all reported series, ranging from 75 to 89% of the cases. It presents as an asymptomatic nodular mass of the posterior lingual midline, usually less than a centimetre in size but sometimes reaching more than 4 cm in size. Larger lesions can present with local symptoms including dysphagia, dysphonia and dyspnoea. Occasionally respiratory difficulty or haemorrhage can occur. At least 15% of patients either present with or are hypothyroid at the time of diagnosis. This patient is unusual in that despite the evidence that hypothyroidism had been present since birth or the early postnatal period, the diagnosis was delayed until the complaint of menorrhagia leading to severe symptomatic anemia led to its detection at the late age of 28. Her lingual thyroid was asymptomatic and was never noticed. A protective, closely knit family environment and inaccessible medical expertise had acted together to prevent earlier recognition and treatment in this case.

Adult patients with lingual thyroid may present with hypothyroidism, however features of cretinism are absent due to adequate thyroid hormone secretion during childhood and thus preventing cretinism and leading to hypothyroidism later in life. Only about 10% of patients with lingual thyroid can present as adult cretin. Hutchison postulated that the age of onset of thyroid insufficiency depends upon the thyroid tissue present. Lingual thyroid is considered as an important cause of sporadic hypothyroidism in children accounting for about 44% cases in one series. Approximately two third of patients with lingual thyroid lack thyroid tissue in neck. In this case ultrasonography neck revealed hypoplastic thyroid tissue but
technetium pertechnate thyroid scan showed no significant uptake in neck, showed a focus of tracer uptake at the root of tongue suggestive of lingual thyroid, as is reported to occur in 70% of cases of lingual thyroid.⁷,¹³,¹⁴

Some have suggested that antibodies directed towards the thyroid cause arrest of descent during early fetal life and poor function.⁵ Gabr proposed that a metabolic defect in thyroid hormone synthesis might be in some way related to the failure of descent.⁶ Alternatively, others believe that since the ectopic tissue is frequently hypoplastic, the maldescent may be secondary to the maldevelopment of the gland.⁷

Thyroid adenoma, goitre, hyperplasia, inflammation and carcinoma can occur in lingual thyroids and need to be evaluated in the same fashion as would any biopsied thyroid gland. Treatment of lingual thyroid has been increasingly conservative in recent years. The current trend is to use thyroid hormone in sufficient dosage to suppress TSH stimulation and minimize goitrous enlargement, which is most likely due to compensatory response to thyroid hypofunction. Surgery is rarely required for complications like cystic degeneration, haemorrhage or suspected malignancy. Treatment with radioactive iodine is no longer routinely used and may induce fibrotic change and scarring.²,¹²

In our patient no local complications were present and the levothyroxine replacement was the only treatment that could be offered to her at this age. Now patient is doing well and is on our follow up.

Conclusion
Early recognition of hypothyroidism is very essential for the normal physical and mental development of an individual and the easy access to medical facilities can bridge the gap between health care providers and health care users. Neonatal screening of hypothyroidism is a ray of hope for early diagnosis of hypothyroidism.

References

Figures

Figure 1