Brief Communication

Juvenile hypothyroidism presenting with hypertrichosis, multicystic ovaries, and pituitary adenoma

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ABSTRACT

Primary hypothyroidism may present with atypical features in children. Here we report a 6-year-old female child with primary hypothyroidism presenting with a combination of several atypical features in the form of hypertrichosis, bilateral cystic ovaries, and feedback pituitary adenoma.

Key words: Hypertrichosis, juvenile hypothyroidism, multicystic ovaries, pituitary adenoma

Introduction

Hypertrichosis and hirsutism are common reasons for seeking medical advice particularly in young children, though only a small proportion of subjects have a diagnosable condition. Hypothyroidism is usually associated with loss of hair. We report a 6-year-old girl with primary hypothyroidism presenting not only with hypertrichosis but other atypical features in the form of bilateral cystic ovaries and pituitary adenoma.

Case Report

A 6-year-old girl was referred for growth of excessive body hair. She was also suffering from poor height gain for 3 years with gradually increasing dryness of skin and lethargy.

She was born at full term, her birth weight was 2.25 kg, and she had normal development milestones and average scholastic performance. She did not have excessive somnolence, cold intolerance, constipation, or headache. There was no history of intake of any medication.

Her height was 104 cm (5th centile target height 159 cm), her weight was 18 kg, and she had normal body proportions. Her pulse rate was 68/min and blood pressure 84/52 mmHg. She had pallor, dry scaly skin and cold extremities. There was a grade1 goiter. Hypertrichosis was particularly noticeable over the lateral aspects of the limbs, the upper back, and forehead [Figure 1]. Her Tanner's sexual maturation score was B1 and P1 for breast and pubic hair, respectively. External genitalia was unambiguously female.

She had normocytic normochromic anemia, hemoglobin 10.8 g/dl (normal 12-14 g/dl). TSH was 146 μIU/ml (normal 0.35-5.5), T_3 42 pg/ml (normal 60-181), T_4 0.6 ng/ml (normal 4.5-12.6). The anti-TPO antibody level was 223.05 IU/ml (normal <5.61 IU/ml) and antithyroglobulin level 135.17 (normal <4.17 IU/ml). Testosterone was 0.11 ng/ml (normal <1 ng/ml), 17-hydroxyprogesterone 0.9 ng/ml (normal <2), dehydroepiandrosterone sulfate (DHEAS) 46 μg/dl (normal 16-96), FSH 0.8 mIU/ml (normal 0.3-2.0), LH 0.39 mIU/ml (normal 0.1-6.0), Prolactin 21.2 μg/l (normal 4.7-23.3).

Radiological investigations revealed a bone age of 4 years (Greulich and Pyle's atlas). Ultrasonography

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of the pelvis showed a prepubertal uterus of size 2.2 × 1.1 × 0.9 cm with bilaterally enlarged multicystic ovaries (right ovary measuring 2.03 × 1.29 cm and left ovary 2.75 × 1.5 cm with increased ovarian volume) [Figure 2]. Magnetic Resource Imaging scan of sella revealed a sellar mass of 1.5 × 1.3 × 1.0 cm size. Technetium 99 m thyroid scan showed poor and patchy uptake of radiotracer suggestive of thyroiditis.

A diagnosis of juvenile hypothyroidism due to autoimmune thyroiditis was made and she was started on a levothyroxine in a dose of 50 μg daily. Dose was uptitrated to 75 μg daily and euthyroid status was established. At 6-month follow-up hypertrichosis had nearly regressed. A repeat USG of pelvic organs showed significant reduction in ovarian size. MRI could not be repeated due to financial constraints.

Discussion

Treatment of hypertrichosis in children is often unsatisfactory as the cause is most often undiagnosed. There is paucity of data on association between hypothyroidism and hypertrichosis in pediatric subset, though congenitally hypothyroid infants often have a low hairline. Perloff reported four cases of what he called hirsutism in children, with body distribution of hair similar to ours, who responded to replacement treatment with thyroid extract.[1] In the case reported by Maekawa *et al.* an underlying abnormality of keratinisation was thought to lead to hair retention.[2] A 10-year-old girl with hypertrichosis and ovarian cysts associated with primary hypothyroidism that resolved with thyroxine therapy has also been described.[3] Nishi *et al.* reported three children with untreated primary hypothyroidism resulting in pituitary hyperplasia and hypertrichosis.[4] These abnormalities disappeared after thyroid replacement as seen in our patient. The pathophysiology of hypertrichosis in our case is difficult to explain. There was no other hormonal abnormality other than severe prolonged primary hypothyroidism; prolactin and adrenal/ovarian androgen were all in the normal range.

Multicystic ovarian disease with hypothyroidism has been previously described in the literature.[5] Various pathophysiologic mechanisms have been proposed, including altered estrogen metabolism, hypothalamo-pituitary axis dysfunction, and altered prolactin metabolism. Ovarian enlargement in the presence of severe hypothyroidism can be due to stimulation of Follicle-stimulating hormone receptors by high TSH levels which is known to have weak FSH-like activity.[6] Increased sensitivity of the ovaries to the circulating gonadotropins could result from the hypothyroid state directly. [7] Ovarian enlargement could also be secondary to a myxedematous infiltration.[8] Our patient too had low gonadotropins with multicystic ovaries, probably due to increased sensitivity of ovaries to gonadotropins or due to myxedematous infiltration of ovaries.

Enlarged pituitary in our case was probably because of thyrotroph hyperplasia due to an end organ deficiency. Enlargement of the pituitary gland or pituitary adenoma have been described in hypothyroidism and as seen in our case some of these patients also had ovarian enlargement and multiple ovarian cysts.[5]

In 1960 Van Wyk and Grumbach described a syndrome of precocious puberty, ovarian enlargement and long-standing juvenile hypothyroidism, commonly autoimmune, undergoing complete regression thyroid hormone replacement. Rastogi *et al.* and Durbin *et al* reported pituitary macroadenoma with Van Wyk and Grumbach syndrome (VWGS).[9,10] Our patient of severe juvenile hypothyroidism of autoimmune etiology presented with a slightly different, reversible syndrome-like presentation of hypertrichosis, multicystic ovaries, and pituitary adenoma; the main difference was that in our case there was no evidence of isosexual precocity; instead there was a very unusual presentation with hypertrichosis.

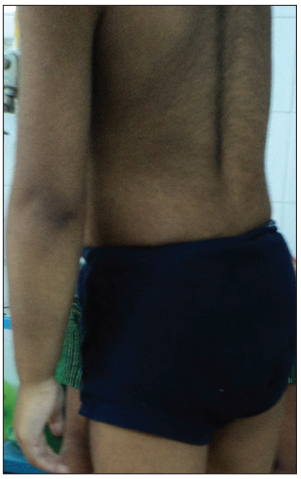

Figure 1: Hypertrichosis

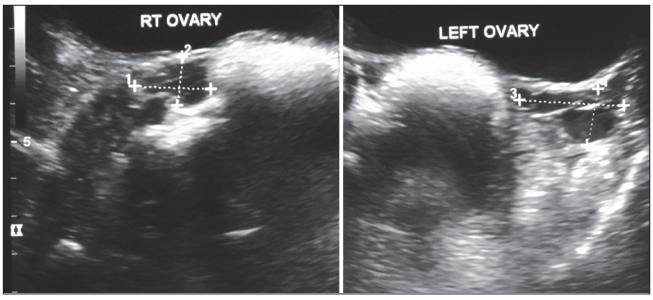

Figure 2: Enlarged multicystic ovaries.

Conclusion

Hypertrichosis, multicystic ovaries, and pituitary adenoma, may rarely be the single presenting feature of severe autoimmune hypothyroidism. Our patient of severe, prolonged, untreated juvenile autoimmune hypothyroidism had all these features in a syndromic presentation, similar but slightly different from VWGS. There may be dramatic regression of hypertrichosis; feedback pituitary adenoma, and multicystic ovaries may also resolve with treatment of severe juvenile hypothyroidsm. Thyroid function should be assessed in all cases of multicystic ovaries, abnormal distribution of body hair, and pituitary adenoma, whether presenting in isolation or in a syndrome like combination.

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