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MULTIPLE ENDOCRINOPATHIES IN A YOUNG FEMALE – A CASE REPORT

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ABSTRACT:

Autoimmune polyendocrine/ polyglandular (APS) syndromes are constellations of multiple endocrine gland insufficiencies, of whom there are 3 types, and amongst which APS type II is the most common. APS type III is defined by the presence of an autoimmune thyroid disease and other autoimmune illnesses, excluding Addison's disease. In our case report here, we present a case of a 22 year old young female from South India with multiple endocrine disorders which come under the category of PGA syndrome type III C, with a few disorders more than the usual endocrinopathies which are included in type III PGA syndromes.

Introduction:

Autoimmune diseases are among the most perplexing of human illnesses.[1] In APS (Autoimmune polyendocrine/ polyglandular syndrome) type III, autoimmune thyroidtis occurs with another organ-specific autoimmune diseases; but the syndrome cannot be classified as APS types I or II due to the absence of Addison's disease. Cases of APS type III associated with different immunological or genetic disorders have been sporadically reported. Premature ovarian failure (POF) is more prevalent than AD (1,000 per 100,000 women), but only 5% of cases are of an autoimmune origin [2,3] In our case report, we are presenting a 22 year old female who has alopecia, primary ovarian failure, dyslipidaemia and bronchial asthma alongside hypothyroidism and type-1 diabetes mellitus. This is another peculiar presentation of the APS type III.

Case report:

A 22 year old female was brought to our casualty unconscious, and was found to be hypoglycaemic which on probing was found that she had been given an improper dose of insulin the previous night. Her glycaemic status was brought to normal soon enough by rushing 25% dextrose, following which she regained consciousness. She was a known case of diabetes mellitus type I under insulin therapy. On obtaining further history it was found that she had hair loss along with failure to attain menarche until now; also with no development of any secondary sexual characters. She happened to be a known hypothyroid under treatment too. She was a twin; and her other twin was a male with normal secondary sexual development and without any significant medical pathology.

On examination, the patient had alopecia with hypertelorism. Otherwise general examination was normal. Systemic examination showed that the patient had bilateral expiratory wheeze and flapping tremor was there initially which later disappeared after one day of admission. On local examination, axillary and pubic hair were absent with absent breast development, Tanner's staging 1.

Furthur investigations showed high fasting and post prandial blood sugar levels, with a high HbA1C, along with significantly high lipid profile with a high total cholesterol, triglyceride and low density lipoprotein (LDL) included. The complete blood count, renal and liver function tests were within the normal limits. ANA was negative serum cortisol levels were within the normal limits. Lutinizing harmone (LH), Follicular stimulation harmone (FSH) levels were sent due to absent secondary sexual characters, and they were found to be high, confirming primary amenorrhea. The thyroid stimulating harmone levels (TSH) was still high, thus confirming inadequate thyroxine dosing; but the microsomal thyroid peroxidase antibody titre was within the normal limits thus excluding the possibility of autoimmune thyroiditis. Ultrasound abdomen revealed the presence of hypoplastic uterus with streak ovaries. Karyotyping was also done suspecting Turner's syndrome, but it turned out to be a normal female geneotype with 46XX. Magnetic resonance imaging (MRI) of the brain was also done which was also normal.

So, with the patient having type I Diabetes mellitus, hypothyroidism, alopecia, primary amenorrhea, dyslipidemia and bronchial asthma, we diagnosed our patient as to be suffering from Autoimmune polyendocrine syndrome type III C. Treatment was given to the patient by correcting the individual abnormalities.

Discussion:

Autoimmune polyendocrine/ polyglandular syndrome (APS) type 3 is an autoimmune condition that affects the body's endocrine glands. The syndrome, which typically affects women during their middle age, results from the failure of the glands to produce their respective hormones. This condition is characterized by autoimmune thyroiditis along with another organ-specific autoimmune disease.[5][6][7] The other autoimmune diseases may include diabetes mellitus, pernicious anemia, vitiligo, alopecia, myasthenia gravis, and Sjogren's syndrome.[6] The adrenal cortex is not involved here. Besides, there are three types of autoimmune polyglandular syndrome type 3[5][6]

- APS3A - Autoimmune thyroiditis with immune-mediated diabetes mellitus (IMDM)
- APS3B - Autoimmune thyroiditis with pernicious anemia
- APS3C - Autoimmune thyroiditis with vitiligo and/or alopecia and/or other organ-specific autoimmune disease

The pathology of the development of APS type 3 has been studied. The underlying autoimmune reaction involves autoantibodies

against endocrine tissues; cell-mediated autoimmunity; or both; and leads to inflammation, lymphocytic infiltration, and partial or complete gland destruction. More than one endocrine glands are involved, although clinical manifestations are not always as expected. The autoimmune reaction and associated immune system dysfunction can also damage non-endocrine tissues.[8]

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