Unusual association of diseases/symptoms

Hypothyroid myopathy mimicking postpolio syndrome

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Correspondence to Professor Rajesh Verma, drrajeshverma32@yahoo.com**Summary**

Hypothyroidism can have diverse neurological manifestations. Myopathy may rarely be the sole manifestation of autoimmune thyroiditis. We hereby report an atypical manifestation of severe hypothyroidism in a middle-aged woman with childhood onset of paralytic polio involving her right leg presenting with a recent onset of increased weakness in the right leg mimicking postpolio syndrome.

BACKGROUND

Hypothyroidism can present with a variety of neurological symptoms ranging from sensorimotor axonal neuropathy, myopathy, cerebellar ataxia, cognitive impairment and depression. Postpolio syndrome (PPS) is suspected in a patient with new-onset weakness or aggravation of preexisting weakness, atrophy, fatigue or muscle pain several years after the episode of acute paralytic polio. However, this is a diagnosis of exclusion. In this case, we highlighted the role of autoimmune hypothyroidism, in aggravating the preexisting weakness due to childhood onset of paralytic polio mimicking as PPS.

CASE PRESENTATION

A 36-year-old woman suffering from childhood onset of poliomyelitis involving the right lower limb having static residual weakness since the last 30 years. During the previous 1 year she started noticing difficulty climbing stairs and buckling of her right knee. The weakness in the right limb progressively increased and since the last 1 month she was unable to stand unsupported which impaired her routine activities and she became bedridden. She had generalised fatigue but denied any history of recent onset of muscle pain, joint pain, fever, cough, skin rash or ulcer. She also denied any symptoms of hypothyroidism such as weight gain, cold intolerance, constipation or symptoms of hyperthyroidism such as weight loss, heat intolerance, frequent bowel movements, tremors or palpitations. She was not on any medications apart from multivitamins and calcium supplements.

On physical examination, her pulse was 72 beats/min, regular, blood pressure was 110/70 mm Hg, body mass index was 26.4. Central nervous system examination revealed normal higher mental function, normal mood and affect with absence of delusions or hallucinations. Cranial nerve examination was normal. On motor system examination, wasting was noted in the right lower limb along with hypotonia (figure 1). Power examination revealed grade 2 power proximally at the hip and grade 3 at the right knee and grade 5 at the ankle joint using MRC (Medical Research Council) grading. The left lower limb revealed grade 5 power as per the MRC grading. Deep tendon jerk at the right knee and ankle was absent.

Figure 1 Patient photograph of the lower limbs demonstrated reduced bulk (atrophy) in right thigh and leg region.

Sensory examination was normal. Rest of the systemic and general examination was normal.

INVESTIGATIONS

Investigations revealed haemoglobin 12.5 g%, total leucocyte count 5200 cells/mm³, platelet count 241 000 cells/mm³, erythrocyte sedimentation rate 21 mm after the 1st hour, blood urea 23 mg/dl, serum sodium 148 meq/dl, serum potassium 4.9 meq/dl, serum ionic calcium 0.98 mmol/dl, random blood glucose 81 mg/dl and liver function tests were within normal limits. The lipid status depicted, total cholesterol 390 mg/dl, triglycerides 150 mg/dl and high-density lipoprotein 48.9 mg/dl. The total creatine phosphokinase (CPK) levels were elevated to 530.7 U/l (24–170). Total triiodothyronine (T3) 0.966 nmol/l (1.3–3.1), total triiodothyronine (T4) 17.25 nmol/l (66–181), thyroid-stimulating hormone >100 µIU/ml (0.27–4.2), anti-thyroid peroxidase titre was raised to 1300 U/ml. The patient was screened for HIV, hepatitis B antigen and hepatitis C, all of which were negative. Antinuclear antibodies by indirect immunofluorescence were negative. Routine cerebrospinal fluid analysis showed five cells, all lymphocytes with normal sugars and protein. Antibodies against cytomegalovirus, Japanese encephalitis virus, herpes simplex virus and

Figure 2 Needle electromyography of right quadriceps suggestive of large-amplitude motor unit action potential and reduced interference.

varicella-zoster virus were not detected. Stool samples sent for the detection of enterovirus and polio virus was negative. Serum cortisol and 25 hydroxy cholecalciferol was found to be normal. MRI thoraco-lumbosacral spine did not reveal any significant abnormality except for thinning of cord. Nerve conduction study showed right-sided common peroneal and tibial nerve axonal neuropathy with normal sensory study. The needle electromyography revealed large-amplitude motor unit action potentials with reduced recruitment in right lower limb muscle suggestive of neurogenic pattern (figure 2). There was no evidence of fasciculations or fibrillations.

TREATMENT

The diagnosis of Hashimoto's thyroiditis with hypothyroid myopathy was established. The patient was started on oral levothyroxine 100 µg/day along with calcium supplements.

OUTCOME AND FOLLOW-UP

The patient started showing improvement within 7 days of starting L-thyroxine and could stand unsupported and after 6 weeks of therapy the patient could walk with help of her brace unsupported and power at hip and knee was grade 4.

DISCUSSION

Recently India has reported no new cases of paralytic polio in 2011.¹ But that is not the end of the story for polio in India as its neighbouring countries (Pakistan and Afghanistan) still have high number of cases and huge number of unvaccinated children. It is equally important to maintain a high vaccination rate and better surveillance so that no new cases are missed. An outbreak of polio was seen in China in 2011, which already declared to be polio free in 1999, due to spread of the polio virus from Pakistan. Despite high awareness, refusal to get children

vaccinated, inadequate surveillance and migrating population poses a great challenge to complete eradication of polio.² However, there is a huge burden of residual paralytic polio cases in our country.

Halstead coined the term postpolio syndrome (PPS) and proposed its diagnostic criteria in 1985 (box 1) and later on revised in 1991.³ An essential criterion of appearance of gradual or abrupt onset of new neurogenic weakness was added in revised criteria of PPS.⁴ Later in 2006, the European Federation of Neurological Societies (EFNS) task force also recommended that the diagnosis of PPS should be based on the revised Halstead criteria.⁵ EFNS task force also emphasised that no test can be diagnostic for PPS and the role of investigations in the evaluation of PPS is to rule out other possible causes of new-onset weakness.

Box 1 Halstead criteria for diagnosis of postpolio syndrome

- ▶ Confirmed history of polio.
- ▶ Partial or fairly complete neurological and functional recovery after the acute episode.
- ▶ Period of at least 15 years with neurological and functional stability.
- ▶ Two or more of the following health problems occurring after the stable period: extensive fatigue, muscle and/or joint pain, new weakness in muscles previously affected or unaffected, new muscle atrophy, functional loss, cold intolerance.
- ▶ No other medical explanation found.

A variety of neuro-muscular disorders such as myelopathy, radiculopathy, endocrine disorders, inflammatory myositis, viral myositis, connective tissue disorders,

inflammatory neuropathy, and so on can worsen the pre-existing weakness in polio patients (box 2).

Box 2 Common differential diagnosis of proximal weakness

- ▶ Cord injury.
- ▶ Motor neuron disease.
- ▶ Lumbar plexopathy (idiopathic, diabetes mellitus).
- ▶ Guillain-Barre syndrome.
- ▶ Chronic inflammatory demyelinating polyneuropathy (idiopathic, paraneoplastic).
- ▶ Congenital muscular dystrophy.
- ▶ Inflammatory myositis such as polymyositis, dermatomyositis, inclusion body myositis, systemic lupus erythematosus, mixed connective tissue disease, scleroderma.
- ▶ Endocrinopathies such as hypo or hyperthyroidism, Cushing's disease, Addison's disease.
- ▶ Vitamin D deficiency, osteomalacia.
- ▶ Infections such as HIV, tropical pyomyositis, West Nile virus, polio, enterovirus and so on.
- ▶ Toxin-induced myopathy such as statin and alcohol.

Hashimoto's thyroiditis, an autoimmune disorder of thyroid gland, is a common cause of hypothyroidism. Hypothyroidism is an endocrine disorder with diverse neurological manifestations ranging from mild cognitive impairment, dementia, cerebellar ataxia, depression, sensorimotor axonal neuropathy, myopathy, and so on.^{6–11} Other autoimmune disorders can coexist along with Hashimoto's thyroiditis such as systemic lupus erythematosus (SLE).¹² Our patient did not have any clinical or laboratory signs of SLE.

Hypothyroidism-induced myopathy is characterised by proximal weakness along with elevation of creatine kinase levels without any correlation with degree of weakness.⁸ Rodolico *et al*¹⁰ reported ten cases of primary autoimmune thyroiditis presenting as isolated persistent myopathy. Similarly in our patient, myopathy was the sole presentation of autoimmune thyroiditis. Occasionally, Hashimoto's thyroiditis can even present with Hoffman's syndrome which characterised by proximal weakness and muscle hypertrophy.¹³ Turker *et al*¹⁴ reported a case of Hashimoto's thyroiditis-induced myopathy with coexisting myaesthetic and Hoffman syndrome-like presentation. The other common causes of proximal weakness are vitamin D deficiency, Cushing syndrome, adrenal dysfunction and pituitary dysfunction.

Apart from poliovirus, West Nile virus (WNV) can cause limb weakness mimicking poliomyelitis. WNV causes acute flaccid paralysis (AFP) due to involvement of anterior horn cells. The recurrent weakness is implicated to persistent infection, excitotoxicity or delayed neuroinvasion.¹⁵ During virological data analysis of AFP surveillance, Coxsackievirus-B, echovirus E6, E11 and E13 were the most frequent non-polio enterovirus isolated from the stool samples.¹⁶ However, our patient responded dramatically to hormone replacement therapy, hence viral infection seems unlikely.

In the setting of preexisting poliomyelitis, the new-onset weakness was more severe in diseased leg mimicking PPS. Mild elevation of CPK levels has also been reported in PPS patients.¹⁷ The asymmetry in the weakness could be explained on the basis of destruction of motor units due to poliomyelitis in the diseased limb along with reduced myosin ATPase activity, calcium release and uptake by sarcoplasmic reticulum and impaired carbohydrate and fat metabolism in the muscle due to hypothyroid state.¹⁸

Nerve conduction studies in hypothyroid patients show sensorimotor axonal neuropathy with patients having predominately sensory symptoms. The needle electromyography study in hypothyroid myopathy is usually normal. Occasionally, mild myopathic pattern or neuropathic pattern may be seen in few patients¹⁹ whereas in PPS there can be presence of fibrillations and fasciculations.

Learning points

- ▶ New-onset weakness, fatigue or muscle atrophy should be thoroughly investigated in known patients of poliomyelitis.
- ▶ Postpolio syndrome (PPS) is a diagnosis of exclusion and should be considered only if no other medical condition can explain symptoms after thorough work-up.
- ▶ Isolated myopathy can be sole manifestation of autoimmune thyroiditis.
- ▶ Creatine phosphokinase levels may be increased in both PPS and hypothyroid myopathy.

Competing interests None.

Patient consent Obtained.

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