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Review Article

STRACHAN-SCOTT SYNDROME: AYURVEDIC PERSPECTIVE

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ABSTRACT

Strachan Scott Syndrome also known as Tropical Ataxic Polyneuropathy (TAN) is a rare neurological disorder characterized by the lesions involving skin and mucous membrane, optic neuropathy and peripheral neuropathy, likely to be caused due to chronic cyanide intoxication following dependency to single plant component, cassava. It is considered as a disease of poor tropical populations but lacks a clear statistics on how many people are affected worldwide, but in affected population prevalence have been noted in more than a quarter. A very little is known about its incidence in India. The clinical spectrum of the syndrome ranges from oculo-oro-genital skin manifestations to severe neurological deficits. This has no direct correlation in classics, though *Kalayakhanja* which is one of the neurological disorders mentioned under *Vatavyadhi,* but clinically different in presentation to Strachan Scott Syndrome. This is a disease caused as a result of chronic intoxication of cyanide so etiologically it could be considered under the concepts of *Dooshivisha*. The signs and symptoms can be considered as the sequential involvement of the *Dhatus* and *Upadhatus*, manifesting as a *Vatavyadhi* as it progresses to involve the nervous system. Being a disease with a very few diagnosed cases and available literatures, this is an attempt in understanding the mechanism of the syndrome from Ayurvedic point of view and the probable management strategies which could be adopted to turn into a manageable one.

INTRODUCTION

The neurological impact of food dependency on plant component is not new to the medical world. Previously it had been reported with the intake of Lathyrus sativa, Lathyrism. Likewise, Strachan Scott Syndrome also known by the name, Tropical ataxic neuropathy, is common in the tropics, which causes significant disabilities as well as heightened mortality and remains unresolved disease with no effective intervention or cure almost since its identification. This was initially reported in Jamaica in 1897 and later the illness was renamed as tropical ataxic neuropathy in 1959. It is characterized by a combination of bilateral optic atrophy, bilateral sensory neural deafness, major posterior column involvement, pyramidal tract myelopathy, and ataxic polyneuropathy.

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The etiology is not clearly identified but existing reports points towards the chronic cyanide intoxication and significant thiamine deficiency due to reliance on a monotonous diet of cassava derivatives[1].

The disease has no direct reference in Ayurveda classics. The concept of *Dooshivisha* may be incorporated leading to sequential manifestation of oculo-oral-genital symptoms and neurological manifestations. The treatment is focused on the *Doshas* involved, *Sthana* and *Lakshanas*. In the initial phase *Pitta Rakta pradhana Tridosha* presentation involving *Aksi-Mukha-Netra-Medra* and later manifesting as *Vata pradhana Tridosha* afflicting *Upadhatu* and presenting as *Sarvangavata*.

Etiopathogenesis

No single factor has been clearly identified.

Chronic Cyanide Intoxication and Tan

A long-standing theory based on the history of nearly complete reliance on a monotonous diet of cassava derivatives in individuals with the syndrome suggests that chronic cyanide toxicity from cassava meals is a major etiological element in TAN. Cyanide is released by the hydrolysis of linamarin, a cyanogenic

glycoside in cassava and the former is detoxified to thiocyanate by the enzyme rhodanese which is excreted in the urine. Patients with TAN have been found to have elevated serum thiocyanate levels [2].

Thiamine Deficiency and Tan

Significant thiamine deficiency was identified with patients with TAN.

a) Firstly, evidence of inadequate dietary thiamine was reported among Nigerians with TAN.
b) Secondly, a total dependence on cassava diet, which contains small amount of thiamine.
c) Lastly, binding of linamarin in cassava with sulfur in thiamine, renders the vitamin inactive by splitting thiamine into pyrimidine and thiazole moieties [2].

Other factors identified include malnutrition, tropical malabsorption, a vegetarian diet, a poor protein intake, viral infection, degenerative and genetic factors [1].

Classification

TAN includes two neurological disorders that can be identified by meticulously reviewing the literature.

1. The first condition is a syndrome that primarily affects adolescents characterized with mucocutaneous signs of malnourishment as well as retrobulbar neuropathy and spinal ataxia in 50%. It is improved with dietary changes and by supplemental of autoclaved dried yeast.
2. The second condition is characterized by different combinations of gait ataxia, ocular atrophy, sensory neural deafness, and sensory polyneuropathy that affect elderly and middle-aged individuals. Unlike the former, malnutrition rarely manifests as mucocutaneous symptoms and dietary changes or vitamin supplements have minimal impact on neurological symptoms [1].

Clinical Manifestation

Skin and mucous membrane lesions, peripheral neuropathy, and optic neuropathy make up the rare neurologic condition, TAN[3]. Typically, a florid, unresolved or fully developed "oculo-oro-genital syndrome" precedes the neurological symptoms [4].

Ocular Manifestation: Fissures in the cornea, accompanied with blepharoconjunctivitis, episcleral infections, and vascularization of the cornea occasionally, associated with symptoms including eyelid sensitivity [4].

Oral Manifestation: There are fissures and rhagades in the mouth overladen by a yellow crust/exudate, which are painful and itchy, and during healing this angular stomatitis becomes depigmented. The early phase is referred to as "sore corners" and the latter stages as "white corners". Cheilosis with epithelial desquamation results in intense burning sensation [4].

Genital Manifestation: The scrotal changes start with intense pruritis, resulting in fine brawny desquamation giving it a shining silvery appearance. Following this fissuring starts resulting in ulcers eventually leading to necrosis of the skin [4].

Neurological Manifestation: Changes starts by involving legs followed by the arms, then the trunk and eventually the mouth and face.

Neurological changes are staged as:

Stage I

Paraesthesia and numbness in the legs often associated with muscular pain and spasms. Both the superficial and deep tendon reflexes are diminished. A slight ataxia occurs while bending or turning. There is numbness and weakness in the hands associated with impaired coordination [4].

Stage II

Characterized by broad base ataxia with a drunken staggering gait associated with features of incoordination. Romberg test is elicited as positive in this stage. Joint position sense is impaired. Diadochokinesia is associated with intensive tremor and dysmetria [4].

Stage III

The patients are unable to get up from sitting position or stand without assistance. The tongue becomes clumsy and speech is impaired. Gustation is also impaired. Both deep and superficial reflexes are abolished without any signs of pyramidal involvement. The intensity of hypoesthesia increases. Movements are painless but associated with numbness all over the body especially on the face. Gradually, this results in a significant degree of asynergy and in coordination in the extremities. The hand movements become athetotic without control. Marked truncal ataxia lead to repeated episodes of falls. The neuropathy advances in a symmetrical manner [4].

Investigations

- Full blood count, (ESR) Erythrocyte sedimentation rate, electrolytes, protein strip, thyroid function, auto-antibody screen including antinuclear factor, rheumatoid factor, anti-DNA antibodies, vitamin B12, red cell folate, red cell transketolase, heavy metal and toxin screen, chest radiograph.
- Lumbar radiogram
- Spinal fluid analysis
- Audiogram
- VER (Visual evoked response) to diagnose problems with the optic nerves and ERG (Electro retinography) to measure the electrical response of light sensitive cells.
- NCS (Nerve Conduction Study) and EMG (Electro myography). [5]

Courses and Prognosis

Patients often experience remissions which require months to fully manifest, and the degree of relief corresponds closely with how severe the initial symptoms were, with less mortality rate [4].

Management

A detailed neurological workup, including CSF analysis and neuroimaging, should be done to patients in order to rule out other disease processes, such as multiple sclerosis. Dietary management is the main form of treatment adopted to combat specific nutritional deficiencies and enhance nutritional status. Early supplementation with Vitamin B-complex and vitamin A supplementation is mostly advised [3].

Ayurvedic Perspective

The explanation which resembles Strachan Scott Syndrome is not accessible as a separate entity in Ayurveda classics. Any disease condition with the involvement of nervous system could be understood under the wider concepts of *Vatavyadhi*. *Vata dosha* has its main *Lakshana* as "*Va gathi gandhanayo*" (*Gathi* - responsible for the bodily movements; *Gandhana* - sensory functions) and hence it is the sustainer of body. This *Vayu* gets vitiated by a) *Dhatu kshaya* and b) *Margasya-avarana* [6].

Nidana* and *Samprapti

The *Nidanas* contributing to the genesis of the disease could be either,

a) *Dhatukshaya*- Caused by malnutrition, poor protein diet, degenerative and genetic factors.

In the early phase of the syndrome, dependency to a monotonous diet is understood as one among the main etiology responsible for TAN, which could be understood as indulgence in *Heena matra ahara* leading to *Vataprakopa*. In Ayurveda classics, references regarding nutritional deficiencies could be considered under *Kuposhanajanya vyadhi* [7]. *Heena matra ahara* is *Indriya upaghatakara* results in *Marma upaghata* [8].

b) *Margaavarodhaja*- Resultant of chronic cyanide intoxication and viral infection (which could be a triggering factor).

Monotonous diet of cassava, i.e., *Atibhojana* of *Ahithabhojana*, results in the production of toxic glucoside in the body as a by-product of impaired metabolism, which can be understood as *Aama*, which persist for an extended period of time, hinders the normal physiological functions of body tissues (*Jataragnimandhya* resulting in circulating *Aama* in the *Srotas* causing *Dhatuvagnimandhya*), which further slows down the metabolism that again contributes to the *Samprapthi* [9].

The toxic glucoside on combining with digestive enzymes hydrolyse to produce cyanide, and the chronic accumulation of cyanide could be understood from the concept of *Dooshivisha* ("*Yat visham puranam*"), initially which remains in a quiescent stage [10]. On continuing *Nidanas* and other triggers including infections causes *Rasadi dhatu dushti* leading to hampered *Dhatvagni* and *Utharothara dhatu dushti*, further to *Sthanasamsraya* in *Upadhatu* and *Marma* resulting in *Vatavyadhi* [11].

Lakshana

Initially, the syndrome commences with oculo-oro-genital symptoms, which could be compared to the involvement of *Dhatus*. Charakacharya has stated that, *Dushivisha* vitiates *Raktadhatu* and causes skin diseases such as *Kitibha* and Kota and then vitiates the *Sareerika doshs* one by one and at last leading to death[12].

Ocular manifestations like blephero-conjuctivitis and episcleral infections can be understood as *Netra abhisynada* caused due to *Majja dhatu pradosha*. Oral symptoms of fissures and epithelial desquamtion as *Asya paka* and scrotal changes as *Medrapaka*, both are the resultant of *Raktha dhatu Pradosha* [11,13].

The involvement of *Dhatu* and *Upadhatu* in the manifestation of disease is enumerated in Table 1[11].

***Dhatu Pradosha Vikaras* and Clinical Manifestation**		
Rasa Dhatu	*Asradha, Aruchi, Gourava, Karshya*	Signs of malnutrition
Rakta Dhatu	*Medra-Asya paka*	Scrotal changes Fissuring of mouth
Mamsa Dhatu	*Oshtaprapaka*	Fissuring over mouth
Majja Dhatu	*Netra abhishyanda*	Bhelpheroconjuctivitis, Episcleral infection
Involvement of *Upadhatus*		
Sira	*Vatavaha Sira*	Neurological manifestations
Twak	*Kushta*	Skin manifestations over mouth (fissuring) and genitalia (Scrotal changes leading to ulceration and necrosis)
Snayu	*Sankocha, Gathi hani, Chesta nasa*	Numbness Loss of motor activities

Further development of neurological symptoms could be understood as the involvement of *Updhatus* as a result of *Sthanasamraya* of the *Dosha* in *Vatavaha Sira* and *Snayu* leading to its *Upasoshana*, manifesting as *Vatavyadhi*. The *Lakshanas* are *Suptata* (numbness), *Swapa* (numbness), *Sada* (*Anganam kriya asamardham*– weakness), *Sanga* (*Vak sanga*– aphasia/ slurred speech) and *Anavasthithathva* (uncoordinated or ataxia symptoms) [14].

The diminution of superficial and deep tendon reflexes and inability to get up from sitting or to stand without support could be understood as *Ceshta nasa* (loss of motor activities), resultant of *Upaghata* to *Siromarma* [15].

Course and Prognosis

In the initial stage, the disease could be controlled by supplementation of autoclaved yeast. As per Susrutha Acharya, *Dooshivisha* is *Sadya*, if within 1 year. Later on, it becomes *Asadhya* [16].

Considering the chronicity, being a *Nithyaanushayitha vyadhi*, manifesting as *Dhatu-Upadhatu pradoshaja* (*Gambeeradhatustha*) and *Marmaasritha* and with involvement of *Dwipada* (*Bahya* and *Madhyama Rogamarga*) it could be considered as *Yapya* [17].

Management

During the initial phase of the syndrome, the general management line of *Dooshivisha* commencing with *Swedhana* followed by *Vamana* and *Virechana* could be done to achieve detoxification of the accumulating toxins [18]. The progressive stage could be managed following the *Vatavyadhi Chikitsa*, based on cause, either *Dhatukshayaja* or *Margaavarodha janya*. Treatment for *Dhatukshayajanya* is *Brhmana* and for *Margavarodhajanya Avastha, Anabhishyandi* and *Vata anulomaka Chikitsa* could be adopted [19,20].

For achieving healing of the ulcerations (*Vrana*), *Upakramas* told in managing *Dusthavrana* could be adopted including *Abhyanthara* and *Bahya shodhana*. *Abhyanthara shodana* could be achieved by *Vamana*, *Virechana*, *Vasti* and *Nasya karma*. *Bahya shodhana* through *Raktamokshana* (*Jalooka avacharana* or *Siravyadhana*), *Vrana prakshalana* and *Vrana pichu dharana* [21].

DISCUSSION

Strachan Syndrome is defined by a combination of cutaneous and neurological symptoms, with oculo-oral-genital manifestations in the acute stage followed by development of neurological symptoms. The specific mechanism of pathogenesis is unknown, but the postulated theory supports to the chronic intoxication due to dependency on a monotonous plant diet, cassava. Dietary supplementation is the management adopted in the initial phases and on progression involving the nervous system, the disease becomes potentially disabling. In Ayurveda any disease with neurological manifestation is understood under the concepts of *Vatavyadhi*. The *Nidanas* contributing to the genesis of the syndrome includes both *Dhatukshaya* (malnutrition/poor protein diet/genetic factors) and *Margavarodhaja* (chronic intoxication/viral infection). Since postulated as a sequalae to chronic cyanide intoxication, the concept of *Dooshivisha* can also be considered. Different modalities of management including the *Chikitsa* principles of *Dooshivisha* in the initial phase, *Vrana Chikitsa* for managing ulcertaions and when advanced with neurological involvement *Vatavyadhi Chikitsa* can be employed.

CONCLUSION

Strachan Syndrome is a rare disease with etiology pointing towards chronic cyanide intoxication due to cassava dependency, which was considered under the concept of *Dooshivisha* and subsequent clinical manifestations to the *Dhatu Prabhava vikaras* as a sequale to *Dooshivisha* ranging from oculo-oral-genital symptoms to severe neurological involvement. In the initial stages of the disease *Dooshivisha Chikitsa* could be adopted, and when progressed to involve the nervous system, *Vatavyadhi Chikitsa* can be adopted.

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