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WILSON DISEASE- A CASE STUDY REPORT

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ABSTRACT
Wilson disease, also known as hepatolenticular degeneration, is a disorder in the liver that results in the improper metabolism of copper, which leads to accumulation of excessive amounts of this vital trace element in the liver, brain, eyes, and other organs. Although copper is essential for normal physiologic function, it can become toxic and life-threatening when too much is present within critical organs, especially the liver and the brain. There is no cure for Wilson disease, and patients affected with this disorder face a lifetime of treatment. Fortunately, the disease is very treatable if diagnosed before significant damage to the liver or brain occurs. This article presents a case study on Wilson disease, where patient was presented with neurological symptoms. This disease was diagnosed as Vatavyadhi and treated as per the concepts of Ayurveda for one month with proper observation. Results of treatment were found satisfactory.

KEYWORDS: Wilson's disease, Copper toxicity, Brain damage, Neurological symptoms, Vatavyadhi.

INTRODUCTION
Wilson's disease, also known as hepatolenticular degeneration and progressive lenticular degeneration, is a rare genetic disorder that causes copper poisoning in the body.[1] It affects about 1 in 30,000 people worldwide.[2] In this disease excess copper stored in various body tissues, particularly the liver, brain, and corneas of the eyes. This causes tissue damage, tissue death, and scarring. If this disease left untreated, it may cause liver (hepatic) disease, central nervous system dysfunction, and death.

Copper plays a key role in the development of healthy nerves, bones, collagen and the skin pigment melanin. Normally, copper is absorbed from our food, and excess is excreted through a substance produced in the liver (bile). But in people with Wilson's disease, copper is not eliminated properly and instead accumulates, possibly to a life-threatening level. Early diagnosis and treatment may prevent serious long-term disability and life threatening complications. Treatment is aimed at reducing the amount of copper that has accumulated in the body and maintaining normal copper levels thereafter.

Signs & Symptoms
Wilson disease is a rare genetic disorder beginning with liver dysfunction where damage begins by six years of age, but usually presents clinically in teenage years or early twenties. Common signs of associated liver disease include a yellow discoloration (jaundice) of the skin, mucous membranes and the membranes (sclera) that line the eye, swelling (edema) of the legs and abdomen (ascites) due to abnormal retention of fluid, presence of abnormal blood vessels in the oesophagus that may bleed (oesophageal varices), a tendency for bruising and prolonged bleeding, and excessive tiredness (fatigue). Some individuals with Wilson disease may have only abnormalities of liver function test and may show no other symptoms until many years later.[2]

In some patients, liver disease does not reveal itself, and the patient develops neurologic (brain-related) symptoms. Common neurological symptoms of Wilson disease that may appear and progress with time include tremor, involuntary movements, difficulty swallowing (dysphagia), difficulty speaking and poor articulation (dysarthria), lack of coordination, spasticity, dystonic postures, and muscle rigidity. Almost all affected individuals with the neurological symptoms of Wilson's disease have Kayser-Fleischer rings in their eyes that can be identified by an ophthalmologist.

The psychiatric manifestations of Wilson disease may vary widely from patient to patient. These symptoms may be confused with other disorders ranging from depression to schizophrenia, and are often misdiagnosed as substance abuse. Changes in personality or behaviour may occur. Most affected individuals with psychiatric symptoms also have neurologic symptoms concurrently or will develop them within about three years and Kayser-Fleischer rings in the corneas of their eyes.

In young females, menstruation may not begin or ceases, until disease is treated. This is due to general disturbances in hormone metabolism due to the liver disease caused by Wilson's disease. Menstrual irregularity, loss of menstruation (amenorrhea), miscarriages and infertility are also common.

Other signs and symptoms of Wilson disease may include kidney stones and renal tubular damage, premature arthritis, and other joint and bone involvement including thinning of the bones (osteoporosis) and the appearance of bony outgrowths (osteophytes) at large joints. There may also be reduced spinal and extremity joint spaces.

Diagnosis

Tests and procedures used to diagnose Wilson's disease include: Blood test and 24 hour urine collection test. Blood tests can monitor liver function and check the level of a protein that binds copper in the blood (ceruloplasmin) and the level of copper in the blood. People with Wilson disease often have low ceruloplasmin levels, but not always. People with Wilson disease may have lower than normal blood copper levels. Urine test will check the amount of copper in urine. Copper levels in the urine are often higher than normal in people who have Wilson disease. If the results of blood and urine tests don't confirm or rule out a diagnosis of Wilson disease, next diagnostic procedure is liver biopsy to check for liver damage and cirrhosis as well as check the amount of copper in the tissue. In people who have nervous system symptoms, imaging tests (CT, MRI) are recommended to check for signs of Wilson disease or other conditions in the brain.[3]

Treatment

Treatment for Wilson disease is life-long and aimed at lowering copper levels to nontoxic levels, and at preventing the progression of the disease and trying to reverse any signs and symptoms that have appeared because of copper accumulation in the body.[4]

Treatment for Wilson disease includes three types of medications. First those that remove (chelate) copper from the body by urinary excretion second, to prevent the gut from absorbing copper from the diet, and third, which both prevents absorbing copper and binds up toxic copper in the blood making it nontoxic. The available copper chelators are dimercaprol, penicillamine, dimercaptopropane sulfonate, and trientine. Zinc administered in the form of zinc salts decreases intestinal dietary copper absorption.

Case study report

Ten years old hindu girl presented to the OPD with symptoms like spasticity, rigidity of muscles, involuntary movements in lower limbs, difficulty in speech, lack of coordination. Symptoms began with deteriorating school performance and increasing lethargy noted over for two years. Recently girl had developed the features of jaundice, taken treatment for the same and got cured. Later gradually she developed present symptoms. Patient was subjected to MRI of brain which revealed bilateral symmetrical T2/FLAIR hypersignal intensities involving bilateral caudate nuclei, thalami, putamen and medial temporatl lobes and bilateral diffuse cerebral atrophy. Examination of eye revealed bilateral Kayser — Fleischer rings but no other abnormality. Serum caeruloplasmin level was <0.10mg/dl and serum copper level was 83.0 µg/dl. Urine volume in 24 hours was 1900ml .24 hours urinary copper excretion was 308.2 microgram per day. Copper urine was 162.21µg/L. Liver biopsy revealed cirrhotic change. She was under copper chelating agent and zinc salts as oral medicine since 6months. No change was observed in her condition. So patient's parents had brought her to our hospital for the further treatment.

Therapeutic Focus

1. Dhanyaka hima 20ml with sugar twice daily in empty stomach for a period1month.
2. Nirocil tablet twice daily with warm water for a period of 1month after food.
3. Pizichil with Ksheerabalataila for 15days
4. Matra basti with Panchatiktaghrita guggulu for 1week.
5. Orally Panchatikta ghrita guggulu 5gms twice daily was started after 1week with hot water.
6. Copper chelating agents and Zinc salts are maintained during the period of treatment.

RESULT

After one month of treatment, the symptoms were started to reduce gradually and she was able to extend both lower limbs and upper limbs completely. Involuntary movements were reduced to maximum extent. Patient was able to speak. Her appetite was good. She was started walk with support. Her urinary copper level came to normal (47.01 µg per 24 h).

DISCUSSION

Copper is an essential metal that is an important cofactor for many proteins. The average diet provides substantial amounts of copper, typically 2-5 mg/day; the recommended intake is 0.9 mg/day. Most dietary copper ends up being excreted. The liver utilizes some copper for metabolic needs, synthesizes and secretes the copper-containing protein ceruloplasmin, and excretes excess copper into bile. Decrease in biliary copper excretion leads to hepatic accumulation of copper. Along with this failure of biliary excretion, there is also reduced incorporation of copper into ceruloplasmin, a serum glycoprotein that contains six copper atoms per molecule and is synthesized predominantly in the liver. When copper accumulates beyond the normal safe storage capacity of the liver, hepatocellular injury results. Furthermore, when the storage capacity of the liver for copper is exceeded or when additional cellular copper is released because of hepatocellular damage, levels of non–ceruloplasmin-bound copper in the circulation are

elevated and copper accumulates in a number of extrahepatic sites, in particular the brain. As copper "spills" over to other organs from the liver, pathologic manifestations become evident in the brain, kidneys, eyes, and joints.

In the brain, the predominant neuropathologic changes of advanced untreated Wilsons Disease are concentrated in the lenticular nuclei, which have the highest copper levels and with increasing copper accumulation undergo various forms of degeneration. Ophthalmologic findings include Kayser-Fleischer rings and sunflower cataracts. Kayser-Fleischer rings are most marked at the upper and lower poles of the limbus, the junction between the cornea and sclera, and are caused by the granular deposition of elemental copper on the inner surface of the cornea in Descemet's membrane. The rings have a golden brown or greenish appearance on slit-lamp examination.

According to Ayurveda, this case of generalized stiffness, and involuntary movements in lower limbs, lack of coordination, difficulty in speech, can correlated to Vatavyadhi, due to agnimandhya at dhatvagni level. The treatment mentioned for Vatavyadhi is snehana, mrudu swedana, virechana and basti.[5] As the patient is weak instead of virechana, matrabasti was given to the patient with Panchatikta ghrita guggulu for a week. As the disease is mainly related to majjavahasrotas, Panchatikta ghrita guggulu is the drug of choice as per the reference of Astanga Samgraha.[6] Externally patient was treated with pizichil of Ksheerabala taila, which gives strength to the muscles.

In Brihat Rasaraja Sundera, the text book of Rasashastra, mentioned antidote for copper toxicity as Munivreehi or Dhanyaka hima along with suger.[7] So along with external treatment, Dhanyaka hima was administered twice daily along with suger in empty stomach. Dhanyaka (Coriandrum sativum L*)* is one of the most useful essential oil bearing spices as well as medicinal plants, belonging to the family Umbelliferae. The seeds of the plant are widely used in folk medicine in addition to its use as a seasoning in food preparation. It is full of antioxidents that demonstrate immune boosting, anti cancer, anti inflammatory and neuro protective effects. The anioxidents in coriander may reduce brain inflammation, improve memory and reduce anxiety symptoms. Reduce unpleasant digestive symptoms like bloating discomfort. It promotes liver functions and bowel movements.[8] Dhanyaka hima is one among the panchavidha kashaya kalpanas mentioned in Sharanghadhara samhita.[9] While explaining effectiveness it is mentioned as dahahara, trishnahara and srotovishodhaka. Same Dhanyaka hima is also said as antidote in copper poisoning. Means through srotoshodhana property it helps in the proper excretion of the deposited copper from the body hence the expected result.

Nirocil tablet was administered twice daily along with warm water. As the biliary system is the major pathway of copper excretion normalising the liver function has got main role in the treatment of Wilson's disease. Hence Nirocil was priscribed. Nirocil each tablet contains 1gm of Bhumyamalaki(Phyllantus niruri), is the best herb is used to normalise the liver function.

After one week of treatment Panchatikta ghrita guggulu was given orally as shamana sneha along with Dhanyaka hima and tablet Nirocil. Panchatikta ghrita guggulu a classical formulations keeps all the three dosha in calm. It is very good blood purifier that helps to eliminate toxins from the body. It has got neuro protective activity as well as it acts as good anti oxident. Also it is good to improve the taste and appetite. Combination of medicines like Panchatikta ghrita guggulu, Dhanyaka hima and Nirocil tablets acts as agnideepaka, srotoshodhaka, raktashodhaka, hence the better result in Wilson diasease a Vatavyadhi. So, it can be concluded that the Ayurvedic approach in such patients may help in providing supportive care and improving the quality of life.

CONCLUSION

A build-up of copper in the central nervous system results in neurological symptoms such as – tremors, uncontrolled movements, difficulty in swallowing, sluggish speech, difficulty in physical co-ordination, muscle stiffness, behavioural changes etc. All these features indicate the involvement of Vatadosha due to dhatvagnimandhya. Classical line of treatment of vatavyadhi along with oral medicines like Panchatikta ghrita guggulu as shamana sneha, Dhanyaka hima as antidote for Copper toxicity and Nirocil tablets as liver protecting agent help in providing supportive care in treating the Wilson disease.

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