A CASE OF METACHROMATIC LEUKODYSTROPHY ASSOCIATED WITH LISSENCEPHALY WHICH RESPONDED TO PANCARKARMA TREATMENT

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ABSTRACT

Metachromatic Leukodystrophy (MLD) is a rare, genetic, degenerative, neurometabolic disorder for which there is no cure. Lissencephaly is a congenital anomaly of the brain where there is absence or less number of gyri. This is a genetic condition hence it is incurable. In this particular case a 1 year and eight month old boy came to the pediatric opd with complaints of not able to sit without support, and not able to stand with/without support, crawl and speech impairment. Examination of central nervous system revealed that speech was affected. The child was able to utter only bisyllables. The motor system was affected. Examination of the upper and lower limb showed reduced muscle power, Grade III. Muscle tone was increased in the left upper limb Grade II. Rest all the limbs the tone and bulk were reduced. Reflexes were sluggish Grade I. Investigations: MRI of the brain was done. It was suggestive of Metachromatic leukodystrophy and lissencephaly. In Ayurveda we can diagnose this condition as amavrita sarvangavata vyadhi. Hence the treatment involves Bahya and Abyanthara snehana(oleation), swedana(sudation), sodhana (purifactory measures) and brihmana treatments and amahara usna thiksha chikitsa. After 17 days of treatment significant improvement was seen in the muscle tone and muscle power. The child also showed improvent in the motor mile stones. The child was able to sit with support and was able to crawl. The child was able to utter 3-4 words like mama, kaka etc. The treatment procedures and the oral medications improved the child’s higher mental functions and motor functions.

KEYWORDS: Metachromatic Leukodystrophy, Lissencephaly, panchakarma treatment, delayed developmental milestones.

INTRODUCTION

Metachromatic Leukodystrophy (MLD) is a rare, genetic, degenerative, neurometabolic disorder that affects approximately one in 40,000 people (primarily children) worldwide. It is an inherited disease, but parents are typically not affected. At present, it is a disease for which there is no cure. The accumulation of sulfatides causes the destruction of myelin (demyelination), which is the protective covering on the nerve fibers that enables communication between the nerves and the brain. The demyelination causes developmental delays, abnormality in the cognitive functions. This is a progressive disorder and has no cure. The condition gets worst and such children have a life span of only 5-10 years. Considering the nature of the disease and clinical symptoms, in ayurveda we can diagnose this case as amavrita saryanga vata vyadhi. Vatavvyadhi management plan can be followed.

Lissencephaly is a congenital anomaly of the brain where there is absence or less number of gyri. This is a genetic condition hence it is incurable. Here also the patients presents with delayed developmental milestones, difficulty in swallowing, abnormal cognitive functions. Only supportive management is available.

CASE PRESENTATION

A boy aged 1 year 8 months came to the pediatric opd with complaints of developmental delay. The boy was not able to sit without support, not able to stand and walk on his own. The other milestones such as social smile, headholding and turning over were achieved but they were delayed. Social smile was acieved at 3-4 months, head holding at 7-8 months, turning over at 5-6 months. There was no h/o fall, trauma or high grade fever, convulsions. The antenatal, birth h/o and post natal h/o were normal.

Examination of higher mental functions revealed that speech was affected. The child was able to utter only bisyllables. The motor system was affected. Examination of the upper and lower limb showed reduced muscle power, Grade III. Muscle tone was increased in the left upper limb Grade II. Rest all the
limbs the tone and bulk were reduced. Reflexes were sluggish Grade I. Investigations: MRI of the brain was done. It was suggestive of Metachromatic leukodystrophy and lissencephaly.

MANAGEMENT AND OUTCOME

Metachromatic Leukodystrophy (MLD) is a rare, genetic, degenerative, neurometabolic disorder. Lissencephaly is a congenital anomaly of the brain and is also a genetic condition hence it is incurable. The patient had complaints of global developmental delay, speech impairment. In Ayurveda we can diagnose this condition as amavrita sarvangavata vyadhii. The treatment of Vatavyadhi includes Bahya and Abyanthara snehana (oleation), swedana(sudation), sodhana (purifactory measures) and brihmana treatments.[1] Since there is involvement of ama dosha, usna thiksha procedures should be done. Hence adwarthana with kolakulathadi choorna was done for 3 days. This was followed by Dhananmala dhara for 7 days. This is also usna and thikshna and hence helps in subsiding ama. Next treatment was to subside vata dosha. For this purpose pizichil (whole body dhara) was done with dhanwanthara thaila for 7 days.

The followings findings were noted before treatment and after treatment.

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<tr>
<th>Sign</th>
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<tr>
<td>Muscle tone</td>
<td>Grade II</td>
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<td>Muscle power</td>
<td>Grade III</td>
<td>Grade IV</td>
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<tr>
<td>Head holding</td>
<td>Achieved</td>
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<tr>
<td>Sitting</td>
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<td>Grade IV</td>
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<td>Standing</td>
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<td>Crawl</td>
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DISCUSSION

In this case study the patient was not able to sit, stand, crawl and walk and speak. The patient had no history of trauma/ high fever/ convulsions. Hence this rules out traumatic causes and infective causes. Birth h/o was also normal. MRI of the brain was done. MRI was suggestive of Metachromatic leukodystrophy and lissencephaly. Considering the above symptoms and investigations we can diagnose this case as sarvanga vatavyadhii. So the treatment would involve vatsavyadhi chikitsa, i.e bahya and abhyantara snehana (oleation therapies), swedana(sudation) sodhana, brihmana procedures[2,3] have to be followed.

First dhananyama dhara was done for 7 days. This procedure is usna and thikshna hence helps in subsiding ama and vata dosha. For the bahya snehana and swedana- pizichil was done using dhanwantaura thaila. pizichil procedure has usna and snigdha gunas and so is vata hara.[4-6] It was done for a period of 7 days. For abhyantara sneha kalyanaka ghrita 1/4 tsp twice daily was given for a period of 20 days. Rajayadi choorna 1/4tsp twice daily was given orally for 20 days. After 17 days of treatment significant improvement was seen in the muscle tone and muscle power. The child also showed improvement in the motor mile stones. The child was able to sit with support and was able to crawl. The child was able to utter 3-4 words like mama, kaka etc. The treatment procedures and the oral medications improved the child’s higher mental functions and motor functions. The treatment has to be continued for next 15 days to achieve the remaining mile stones. Further more the treatment has to be repeated every 3 months to get good improvement in the child.

CONCLUSION

1. Metachromatic Leucodystrophy presenting with delayed developmental milestones can be co related with amavrita sarvanga vatsavyadhii.
2. Dhananyama dhara helps in subsidng the prakupita kapha and vata.
3. Bahya and abhyantara snehana help in subsiding vata and hence helps in improving the motor functions.
4. Abyanthara snehana using kalyanaka ghrita helps in nourishing the mastishkhagata majja and also in subsiding vata and hence improves the higher mental functions.
5. The treatment should be done once in every 3months to get complete relief.

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