Case Report

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APPLICATION OF *KSHARASUTRA* OVER SURGICAL EXCISION OF NEUROFIBROMA (NF1):

A CASE REPORT

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ABSTRACT

Neurofibromatosis type 1 (NF1) or Von Recklinghausen's disease is a rare genetic disorder characterized by the development of multiple noncancerous tumors of nerves and skin (neurofibromas). Head and neck neurofibroma are generally located in the soft tissue. Here, a case of a 42-year-old man with NF1 has been presented. The disease started in childhood with the appearance of multiple hyperpigmented skin macules. He has generalized freckling and café-au-lait spots all over the body and cutaneous nodules with multiple sizes. The diagnosis of NF1 was made according to presence of two or more diagnostic criteria of the National Institute of Health Consensus Development Conference. On the basis of symptoms, it was closely related to the *Kaphaja Arbuda*. *Acharya Sushruta* mentioned *Kshara* and *Ksharasutra* in *Arbuda Chikitsa*. So, *Ksharasutra* was tied on the root of one of the neurofibroma size of 4 x 2.5 cm present on medial side of the mid arm. The Fibroma was shed-off completely after 7 days and no recurrence was observed in a follow up at the end of 12 months after application of *Ksharasutra*.

Keywords: *Arbuda*, Ayurveda, Café-au-lait, *Ksharasutra*, Lisch nodule, Neurofibroma,

INTRODUCTION

The term neurofibromatosis (NF) is used for a group of genetic disorders that primarily affect the cell growth of neural tissues. There are two forms of NF: Type 1 (NF1) and type 2 (NF2)¹⁻³. These two forms have few common features and are caused by mutations on different chromosomes i.e., 17th and 22nd respectively^{4,5}. NF1 also known as Von Recklinghausen's disease, is the most common type of neurofibromatosis and accounts for about 90% of all cases. It is one of the most frequent human genetic diseases, with a prevalence of one case in 3,000 births. There is no sex or racial predilection. It is an autosomal dominant disorder with a basic defect in the embryonic neural crest cells that give rise to ectodermal and mesodermal derivatives⁶ which caused by a spectrum of mutations affecting the NF1 gene located at the 17q11.2 chromosome. NF1 can be of subcutaneous, Cutaneous or plexiform neurofibromas. Only 50% of the NF1 patients have a positive family history of the disease. Café-au-lait spots, axillary and inguinal freckling, optic gliomas, Lisch nodules (pigmented hamartomas of the iris), spinal and peripheral nerve neurofibromas, neurological or cognitive impairment, scoliosis, abnormalities in the oral and maxillofacial region, malignant tumors of the nerve sheath, pheochromocytoma, vasculopathy, and specific bone lesions are common clinical features of NF1.^{2,5} Oral manifestations can be found in almost 72% of NF1 patients.⁷ Malignant progression is generally considered the main cause of mortality, occurring in 2% to 16 % of cases⁸. Histologically, neurofibromas are composed of a mixture of Schwann cells, perineurial cells, and endoneurial fibroblasts, which are not capsulated.⁹ Schwann cells account for about 36% to 80% of the lesion cells.

Clinical diagnostic criteria of Neurofibromatosis-1 (NF1)

Patients have two or more of the following symptoms-

- 6 or more café-au-lait macules (> 0.5 cm in children of > 1.5 cm in adults)
- 2 or more cutaneous or subcutaneous neurofibromas or one plexiform neurofibroma
- Axillary or groin freckling
- Optic glioma
- 2 or more Lisch nodules (iris hamartomas visualized on slit lamp examination)
- Sphenoid wing dysplasia or bowing of long bone (with or without pseudarthrosis)
- First degree relative with an NF1 diagnosis¹⁰

Review of *Ksharasutra*

Ksharasutra therapy, a minimal invasive parasurgical procedure and time-tested therapy has been described in Ayurveda by *Sushruta*, *Charaka* and *Vagbhata*¹¹. *Acharya Sushruta* has described *Ksharasutra* application in the treatment of *Arbuda*¹².

Preparation of *Ksharasutra*

The Standard *Ksharasutra* is prepared by 11 coatings of *Snuhi Kshar* (*Euphorbia nerifolia*), 7 coatings of *Snuhi* latex and *Apamarga Kshara* (*Achyranthes aspera*) and last 3 coatings of *Snuhi* latex and *Haridra Churna* (*Curcuma longum*). The pH of *Apamarga Kshara Sutra* is 9.72¹¹.

Mechanism of action of *Ksharsutra*

- Chemical cauterization and mechanical strangulation of the mass.
- Local gangrene of the tissue mass.
- Ischemic, necrosis and debridement of unhealthy tissue.
- Sloughing of the mass within 5-7 days.
- Healing of the resulting wound takes 10-15 days.¹¹

Advantages of *Ksharsutra* therapy

- Simple and safe of parasurgical procedure.
- Cost-effective.
- Minimal recurrence rate.
- No surgical complications like incontinence, stenosis and stricture.¹¹

Case Report

A 42-year-old man was presented to OPD at National institute of Ayurveda on dated 20th December 2019. He has generalized freckling, café-au-lait spots all over the body and cutaneous nodules NF1 with multiple sizes (Figure A). The disease started in his childhood with the appearance of multiple hyper-pigmented skin maculae. He has slight pain on whole body particularly on fibroma sites.

Consent

The patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand

that his name and initials will not be published, and due efforts will be made to conceal his identity.

Family history

There was not any relevant family history.

Physical examination

Dermatological status

Hundreds of Cutaneous nodules were presented on all over the body; multiple café-au-lait spots with diameter > 1.5 cm; axillary and inguinal freckling. The mucous membranes were not affected.

Ophthalmological status

There were no Lisch's nodules on the iris of both the eyes.

Procedure

Following all the pre-surgical measures *Ksharsutra* was tied on the root of neurofibroma present on the medial side of right mid arm. Turmeric powder was applied on the wound for two days after shedding-off neurofibroma.

RESULT

Neurofibroma was completely shed off by the application of *Ksharsutra* after 7 days and no recurrence was observed in a follow-up after 12 months.

Figure A. Showing generalized neurofibromatosis (NF1) on both upper limbs.
Figure B. *Ksharasutra* tying on the root of neurofibroma on medial side of the right mid arm.
Figure C. Scar after shedding of neurofibroma.

DISCUSSION

Diagnosis of NF1 is usually achieved when 2 or more criteria, developed by the NIH, are met.¹⁰ Neurofibromas are generally diagnosed clinically with appreciation of the typical features, and histopathology is useful to exclude malignant transformation.¹³ In conventional medical science there is no treatment for neurofibromatosis. Usually for neurofibromas resecting surgery is done, aiming at deforming masses and cancerous tissue when malignant transformation occurs. However, these masses tend to recur in 20%.¹⁴

According to Principle of *Ayurveda*, there is no disease without the derangement of *Doshas*. *Acharya Sushruta* has described *Arbuda* as "The *Dosha* having vitiated in any part of the body and

afflicting the *Mamsa*, and produce a swelling, which is circular, fixed, slightly pain, big in size, broad based, slowly growing and does not suppurate." *Sushruta* has classified disease *Arbuda* in to six types mainly *Vataja Arbuda*, *Pittaja Arbuda*, *Kaphaja Arbuda*, *Raktaja Arbuda*, *Mamsja Arbuda* and *Medoja Arbuda*. Among its type *Kaphaja Arbuda* has sign and symptoms of skin colored, stony hard and slow growing, Swelling, less painful, itching¹⁵ which is closely related to neurofibroma of the present case. Surgical removal, *Agnikarma* and *Ksharakarma* are the treatment procedures along with specially indicated medicines. *Acharya Sushruta* mentioned the use of *kshara* in *Kaphaja Arbuda*¹⁶ also described to treat *Arbuda* by tying *Ksharasutra* at the root of *Arbuda*.¹⁷ *Ksharakarma* is better than excision as it has the properties of *Tridoshaghna*, *Dahana*, *Pachana*, *Daran* and *Ropan*.¹⁸ *Ksharasutra* works by chemical cauterization and

mechanical strangulation of the mass following by local gangrene of the tissue mass and ischemic, necrosis and debridement of unhealthy tissue. Sloughing of the mass within 5-7 days and healing of the resulting wound takes 10-15 days. On basis of classical symptoms of *Kaphaja Arbuda* is closely related to the present case neurofibroma. So, in this present case *Ksharasutra* is applied to the neurofibroma (*Arbuda*) on the medial side of right mid arm. It was observed that there was no recurrence in the follow-up at the end of 12 months.

CONCLUSION

In neurofibromatosis (NF1), *Ksharasutra* can be applied for excision of neurofibroma to avoid its recurrence.

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