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PENIS: A RARE SITE FOR NEUROFIBROMA

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ABSTRACT
Neurofibromatosis (NF) comprises a distinct genetic disorder characterized by benign growth of peripheral nerve sheaths, neurofibromata and café au lait macules (CALM), associated with various other cutaneous and systemic manifestations. A patient may present with thousands of neurofibromata over the skin, however, genitourinary neurofibromas in neurofibromatosis type 1 are rare and the neurofibromas involving genitalia in males are even more infrequent than neurofibromas with clitoral involvement in females. Herein, we present a 10 years old child with histopathologically proved penile neurofibroma in addition to other lesions.

KEYWORDS: neurofibroma, CALM, penile.

INTRODUCTION
The neurofibromatosis comprise several distinct genetic disorders that lead to the formation of tumours surrounding nerves and many others pathological features. The two main forms are neurofibromatosis 1 (NF 1) and neurofibromatosis 2 (NF 2), although eight clinical types have been described by Ricardi (Table 1).[1]

Table -1 clinical types of NF.

NF1	Von Recklinghausen's disease
NF2	Acoustic
NF3	Mixed (multiple brain & spinal tumours with CALM & neurofibroma)
NF4	Variant (both CALM & neurofibroma are present but further categorization is not possible)
NF5	Segmental (CALM and/ or neurofibroma is limited to a unilateral segment; non familial)
NF6	CALM (multiple CALM without neurofibroma; familial/ sporadic)
NF7	Late onset (manifestations after 20 years of age; CALM may be absent)
NF8	Not otherwise specified (definite NF, but not characteristic of any other category; CALM may be absent)

NF 1 is an autosomal dominant disorder that affects approximately 1 in 3000 individuals. It is characterized clinically by café-au-lait macules, neurofibromas, freckling in the axillae and groin, Lisch nodules and bony defects.

Diagnostic criteria for NF 1 include seven features, out of which two or more should be present to diagnose a case as NF 1. In its fullest expession, NF1can manifest as thousands of neurofibromas in a patient, hence the appellation neurofibromatosis.
However, penile neurofibromas are reported rarely. Here in, we report a 10 years old male child with a penile neurofibroma in addition to other lesions.

CASE REPORT
A 10 years old male child presented with multiple brown coloured asymptomatic patches over the body along with two skin coloured, raised, asymptomatic, soft to rubbery lesions. One of these soft, rubbery lesions was present

over the back and another one was present on the dorsal aspect of the penis. These lesions were gradually increasing in size and the lesion over the penis was leading to penile deformity. There was no history of any other systemic complaint.

There was a history of presence of similar lesions in the father and grandfather.

Clinical examination revealed presence of multiple tan to dark brown macules ranging in size from 1x1 cm to 3x3 cm over the body (fig 1).

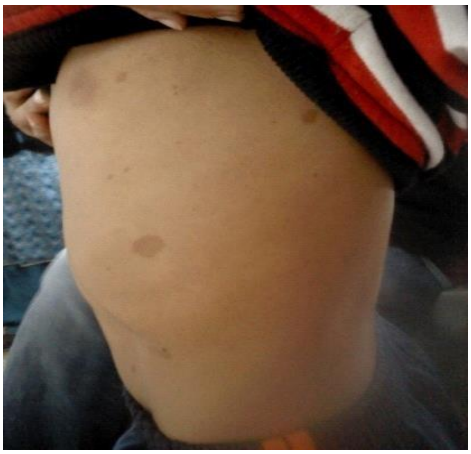

Fig. 1

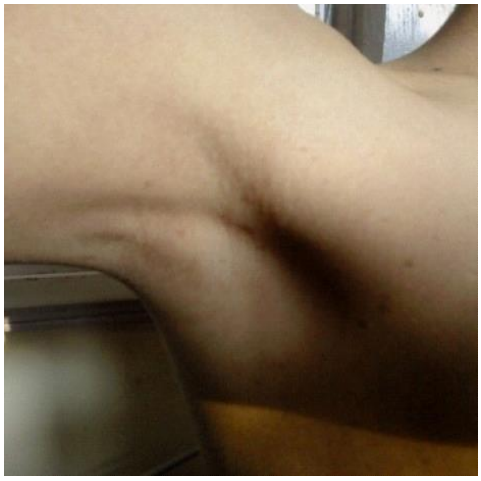

Fig. 2a

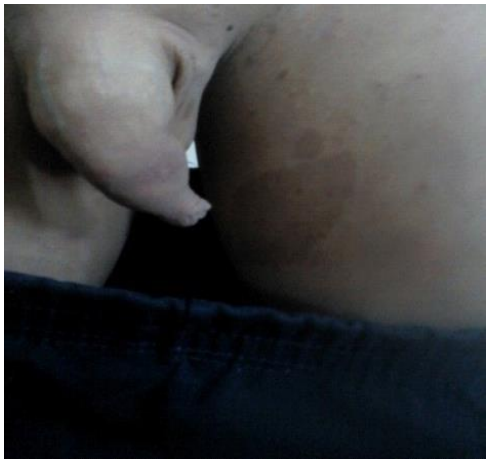

Fig. 2b

Axillary and inguinal freckling were present (fig 2a & 2b). There was a smooth, well defined, skin coloured nodulo-plaque lesion of size 2x2 cm over the right infrascapular region of the back (fig 3).

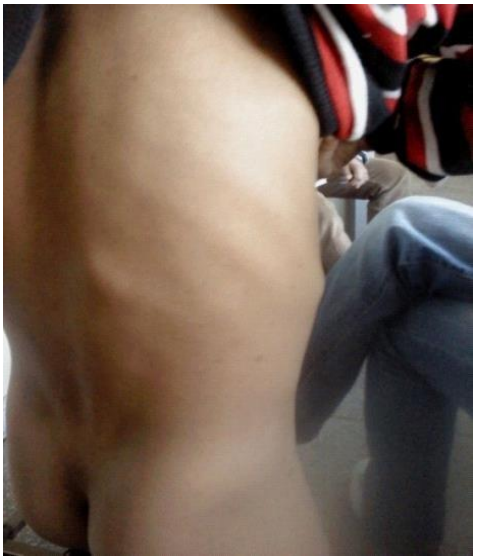

Fig. 3

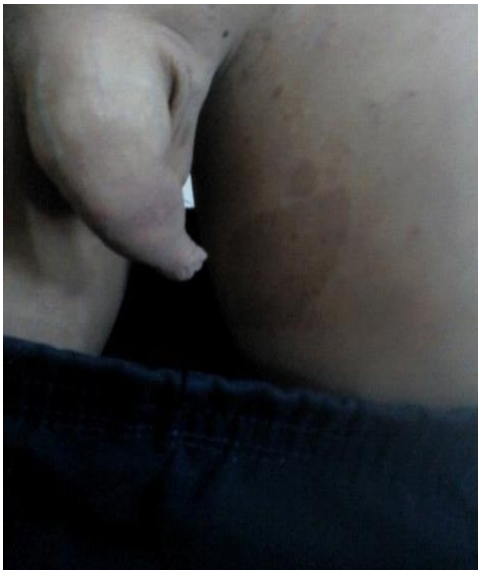

Fig. 4

Also, over the dorsum of penis, there was a skin coloured nodulo-plaque lesion of size 3x4 cms (fig 4) was present. Both these nodulo-plaque lesions were rubbery in consistency and were non tender on palpation. On eye examination, there were lisch nodules present on bilateral iris (4 in number).

Laboratory examination of blood (complete haemogram and biochemical parameters) were within the normal limits. MRI pelvis/ perianal region gave a possibility of penile neurofibroma (fig 5).

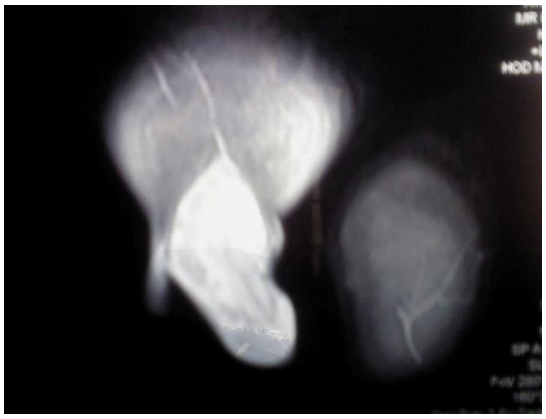

Fig. 5

Surgical excision of penile lesion was done in the department of surgery, IGMC, Shimla (fig. 6,7) and the tissue was sent for histopathological examination. Histopathological examination of removed tissue confirmed neurofibroma (fig. 8).

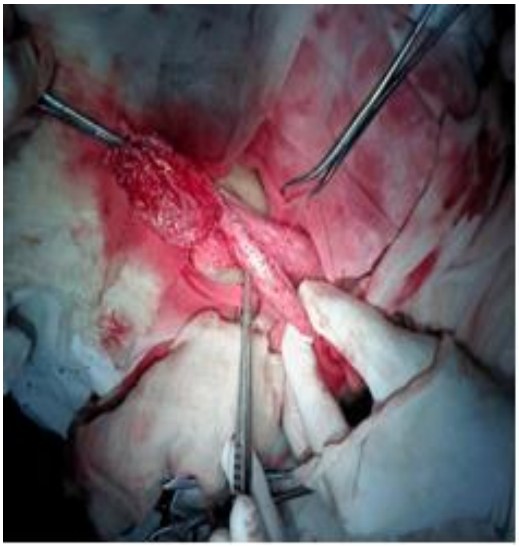

Fig. 6

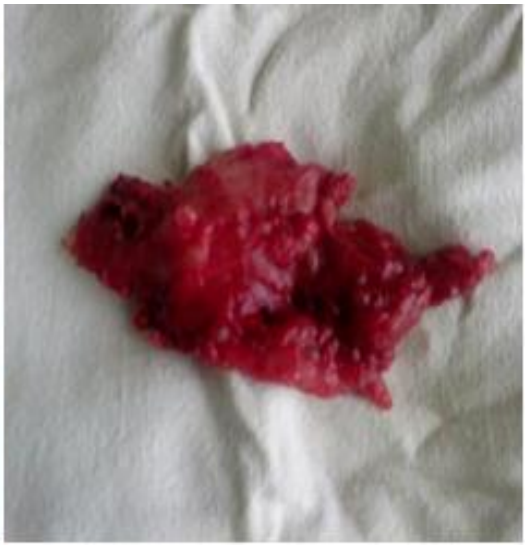

Fig. 7

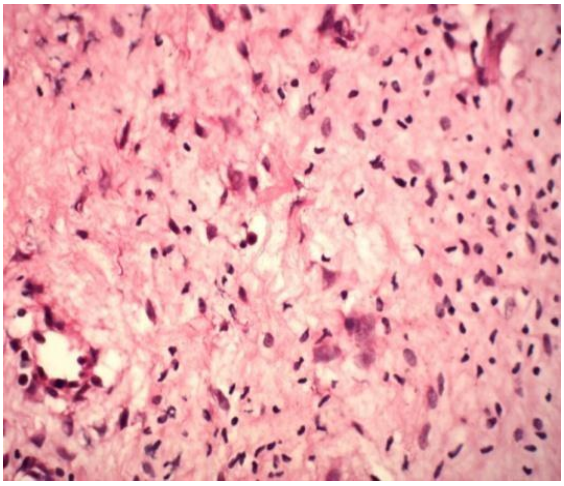

Fig. 8

DISCUSSION

Neurofibromatosis is a neurocutaneous disorder characterized by cutaneous lesions as well as peripheral or central nervous system neoplasms. In 1882, Friedrich von Recklinghausen described this disease pointing out that the skin tumours were derived from peripheral nerves.[2] In retrospect, Virchow first reported a family with more than one affected member.[2] It is now recognized that NF1 is an inherited neuroectodermal abnormality, characterized by benign growths of peripheral nerve sheaths, neurofibroma and café-au-lait macules (CALM), associated with various other cutaneous and systemic manifestations.

The mode of inheritance is autosomal dominant, with almost 100% penetrance by the age of 5 years. Sporadic cases result from a high gene mutation rate and account for up to 50% of all cases. The prevalence of NF1 has been estimated at a frequency of 1 in 2500–3300 births. Incomplete or monosymptomatic forms are frequent. The gene for NF1 is located on chromosome 17. The NF*1* gene has now been cloned and encodes a protein named neurofibromin. The National Institutes of Health Consensus Development Conference Statement, developed in 1988, has proved to be very useful and is widely employed in clinical practice. This requires two or more of the following criteria to be fulfilled.[2]

1. Six or more café-au-lait macules of over 5 mm in greatest diameter in prepubertal individuals and over 15 mm in greatest diameter in postpubertal individuals
2. Two or more neurofibromas of any type or one plexiform neurofi broma
3. Freckling in the axillary or inguinal regions
4. Optic glioma
5. Two or more Lisch nodules
6. A distinctive osseous lesion such as sphenoid dysplasia or thinning of the long bone cortex with or without pseudoarthrosis
7. A first-degree relative (parent, sibling, offspring) with NF1 by the above criteria.

Although it is not difficult to diagnose a neurofibroma, in difficult cases biopsy can be diagnostic.

There is no treatment for the disease, but small cutaneous or subcutaneous neurofibromas can be removed if they are painful.

Solitary involvement of the genitourinary tract by von Recklinghousen disease (VRD) is extremely rare.[3,6] It leads to cosmetic deformity and sexual dysfunction.

The treatment for penile mass consists of sub coronal incision and resection of the lesion. In case of plexiform neurofibroma compromising penile function, total resection of penile mass offers a good therapeutic option. The treatment goal is complete resection with preserving the neurovascular bundle to prevent recurrence or malignant degeneration.[4,5,6]

Even though neurofibromas occur all over the body, they show definite proclivity for developing in the nipple and areola particularly in a female.[7] The penis, however, is a very rare site and hence the occurrence of penile neurofibroma is reported herein.

REFERENCES

1. Arun C Inamdar, Aparna Palit. Genodermatosis. In: R. G. Valia, Ameet R Valia, editors. IADVL Textbook of Dermatology. 3rd ed. Mumbai, India: Bhalani Publishing House, 2008; 133.
2. Irvine AD, Mellerio. Genetics and Genodermatoses. In: Burns T, Breathnach S Cox N, Griffiths C, editors. Rook's Textbook of Dermatology. 8th ed. Singapore: Wiley Blackwell, 2010; 15.15-18.
3. Kousseff BG, Hoover DL. Penile neurofibromas. Am J Med Genet. 1999 Nov 5; 87(1): 1- 5.
4. Barros R, Cavalcanti AG, Favorito LA. Plexiform Neurofibroma with compromising of the penile function. Urology, 2008 Mar; 71(3): 546e9-10.
5. Littlejohn JO, Belmass AB, Selby D. Plexiform Neurofibroma of the penis in a child. Urology, 2000; 56(4): 669.
6. Garaffa G, Biltocehi C, Christopher N, Ralph D. Plexiform Neurofibroma of penis associated with erectile dysfunction due to arterial stealing. J Sex Med, 2008 Jan; 5(1): 234-Epub2007oct24.
7. Sherman JE, Smith JE. Neurofibroma of breast and nipple areola area. Ann Plast Surg, 1981; 7: 302-7.