CASE REPORT

Plexiform Neurofibroma: A Rare Case Scenario

Swati Shandilya

Department of Dermatology, Smt. Kashibai Navale Medical College and General Hospital, Pune, Maharashtra, India

ABSTRACT

Neurofibromatosis Type 1 has a rare variation called plexiform neurofibromas (30%). Multiple nerves give rise to neurofibromas, which appear as oblong, deformed lumps made of connective tissue and skin folds. Most diagnoses are made clinically. Surgery is the main form of proprioceptive neuromuscular facilitation (PNF) treatment. Because PNF can be big and transcend tissue boundaries, removal is challenging. Presenting our experience of such rare case visiting our center.

Key words: Plexiform Neurofibroma, PNF, MRI

INTRODUCTION

Plexiform neurofibromas, an uncommon variety of Type 1 neurofibromatosis, account for 30% of cases. Neurofibromas are oblong, malformed lumps consisting of connective tissue and skin folds that are caused by many nerves. The majority of diagnoses are done clinically. Proprioceptive neuromuscular facilitation (PNF) is mostly treated by surgery. PNF can be large and cross tissue borders, making removal difficult. Here, an unusual case of a 22-year-old man who visited with a progressive facial deformity that began in childhood is presented.

CASE REPORT

A 22-year-old man with a developing facial abnormality that started in childhood was seen.

On the trunk and arms, a skin examination also found numerous neurofibromas, freckles, and café-au-lait macules, as well as numerous nodules with center depressions known as "buttonhole" signs that were non-tender and unrelated to any other symptoms.

Slit lamp evaluation revealed characteristic Lisch nodules on the iris.

History revealed similar complaints in grandfather.

Access this article online	
Quick Response code	**Website:** www.mjbar.in
	Recevied on: 16/12/2022 **Accepted on:** 19/01/2023

RESULTS

On Examination

Non-tender mass was noted in the left as well as the right temporal region. He was investigated as follows:

Magnetic resonance imaging (MRI) head-and-neck-large lobulated mass in the left periorbital soft tissues and adjoining left frontotemporal scalp tissues giving homolateral eye dislocation s/o plexiform neurofibroma.

Ocular motility was affected.

On Histopathology

The biopsy shows a well to ill circumscribed non-epithelial neoplasm made up of oval and spindle shape cells and filling up most of the dermis.

Overlying papillary dermis and few appendages were spared.

He was diagnosed as neurofibromatosis Type 1 (NF-1) on the basis of histopathology and clinical examination.[1,2]

He underwent surgical excision of neurofibroma under GA. In post-surgery, there was recurrence of mass growth for

Address for correspondence:
Dr. Swati Shandilya, Senior Resident, Department of Dermatology, Smt. Kashibai Navale Medical College and General Hospital, Pune, Maharashtra, India. Mobile: +91-7020358436.
E-mail: shandilyaswati93@gmail.com

Figure 1: Plexiform mass

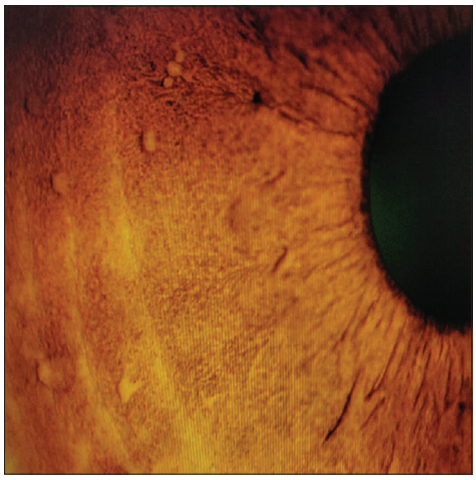

Figure 2: Slit lamp examination showing lisch nodules

which he preferred to be under conservative management [Figures 1 and 2].

DISCUSSION

Plexiform neurofibromas are a rare form of NF-1 in which connective tissue and skin folds are also affected as well as bulging and deforming masses that develop from numerous nerves. Lesions are described clinically as "bags of worms."

Up to 30% of NF-1 individuals include plexiform neurofibromas, which are particularly common in the craniomaxillofacial area.

The primary signs include:

Eye involvement dyspnea causes respiratory failure and upper airway tightness causes abnormalities in eye motility.

Neurological issues brought on by cranial nerve compression.

Due to facial deformities, social isolation, sadness, and other mood problems.

Diagnostic Criteria for NF-1

a) 6 or more café au lait macules (>0.5 cm in children or >1.5 cm in adults)
b) 2 or more cutaneous/subcutaneous neurofibromas or one plexiform neurofibroma
c) Axillary or groin freckling
d) Optic pathway glioma
e) 2 or more Lisch nodules (iris hamartomas seen on slit lamp examination)
f) Bony dysplasia (sphenoid wing dysplasia, bowing of long bone±pseudarthrosis)
g) First degree relative with NF-1.

Complications

Neurological, cosmetic, and other clinical impairments may result from plexiform neurofibroma.

Plexiform neurofibromas may develop in specific locations and result in serious clinical consequences[3]

A malignant peripheral nerve sheath tumor develops from about 10% of plexiform neurofibromas[4]

Treatment

Surgery is the main course of treatment for plexiform neurofibroma.

While resecting invasive neurofibromas in the head-and-neck region, extensive surgical procedures must be evaluated against functional impairments that are almost certain to occur.

Radiation and chemotherapy can be used to treat plexiform neurofibromas once they have turned malignant.

For timely discovery and a repeat operation to achieve additional correction, periodic clinical examination and MRI evaluation are necessary for roughly 2 years.

CONCLUSION

We offer a case study of the incredibly uncommon condition known as plexiform neurofibroma.

Due to the extreme facial deformities brought on by the disease's course, normal life was restricted.

Proprioceptive neuromuscular facilitation (PNF) provides conclusive evidence for NF-I diagnosis.

Finding the local infiltration and detailed architecture of tumors in the head-and-neck region with MRI can be helpful.

To rule out recurrence or malignant conversion, patients with PNF should be frequently called back.

REFERENCES

1. Neurofibromatosis. Natl Inst Health Consens Dev Conf Consens Statement 1987;6:1-7.
2. Packer RJ, Gutmann DH, Rubenstein A, Viskochil D, Zimmerman RA, Vezina G, *et al*. Plexiform neurofibromas in NF1: Toward biologic-based therapy. Neurology 2002;58:1461-70.
3. Wise JB, Patel SG, Shah JP. Management issues in massive pediatric facial plexiform neurofibroma with neurofibromatosis Type 1. Head Neck 2002;24:207-11.
4. Isler MH, Fogaça MF, Mankin HJ. Radiation induced malignant schwannoma arising in a neurofibroma. Clin Orthop Relat Res 1996;325:251-5.

How to cite this article: Shandilya S. Plexiform Neurofibroma: A Rare Case Scenario. Med J Basic Appl Res 2023; 4(1):35-37.

Conflicts of Interest: None. **Source of support:** None.