A rare case report of Solitary Forehead Neurofibroma in a 19-Year-Old Female

Dr. Jugajyoti Pathi, Dr. Vikas Jha, Dr Ahsan Abdullah, Dr Meghana S Patil, Dr. Anita Mohanty, Dr. Malvika Raghuvanshi

ABSTRACT

Background

Neurofibromas are benign peripheral nerve sheath tumors that may present as solitary lesions or as part of neurofibromatosis type 1 (NF1). Solitary neurofibromas of the forehead are uncommon and can be mistaken for other soft tissue masses, delaying diagnosis and treatment.

Case Presentation

A 19-year-old female presented with a painless, slowly enlarging left forehead swelling since four years. Clinical examination revealed a 3 X 2 cm, firm, mobile, non-tender subcutaneous mass with intact overlying skin. Excisional biopsy was performed; histopathology confirmed a neurofibroma. The patient had an uneventful recovery and remained free of recurrence after 12 months.

Conclusion

Solitary forehead neurofibroma should be included in the differential diagnosis of chronic forehead swellings. Accurate preoperative assessment, careful surgical planning to preserve cosmesis, function and a histopathological confirmation are essential. Long-term follow-up is recommended to detect recurrence or new lesions.

Keywords

Neurofibroma, Forehead swelling, Solitary peripheral nerve sheath tumor, Excisional biopsy

INTRODUCTION

Neurofibroma is a benign tumor of peripheral nerve sheath origin that is composed of a mixture of Schwann cells, perineurial-like cells, fibroblasts, and mast cells embedded within a myxoid stroma **[1]**. Neurofibromas may occur as solitary sporadic lesions or as part of Neurofibromatosis Type 1 **[2,3]**, an autosomal dominant disorder caused by pathogenic variants in the NF1 gene. Head and neck involvement accounts for a substantial proportion of neurofibromas, but isolated lesions of the forehead are relatively uncommon **[4]**.

Approximately 90% of neurofibromas occur sporadically, while the remaining cases are associated with Neurofibromatosis Type 1 or

1. Dr. Jugajyoti Pathi, Professor, Department of Oral & Maxillofacial Surgery, Kalinga Institute of Dental Sciences, KIIT Deemed to be University, Bhubaneswar. Email Id – jpathi@kims.ac.in
2. Dr. Vikas Jha, Lecturer, Department of Oral & Maxillofacial Surgery, Kalinga Institute of Dental Sciences, KIIT Deemed to be University, Bhubaneswar. Email Id - jha.vikash980@gmail.com
3. Dr Ahsan Abdullah, Professor, Department of Paediatric and Preventive Dentistry, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow. Email: klahsan@gmail.com
4. Dr Meghana S Patil, Senior Lecturer, Department of Oral and Maxillofacial Surgery, Dayanand Sagar College of Dental Sciences, Bangalore. Email id: meghanapatil1997@gmail.com
5. Dr. Anita Mohanty, Lecturer, Department of Oral & Maxillofacial Surgery, Kalinga Institute of Dental Sciences, KIIT Deemed to be University, Bhubaneswar. Email Id – dranitamohanty07@gmail.com
6. Dr. Malvika Raghuvanshi, Professor, Department of Oral & Maxillofacial Pathology, Kalinga Institute of Dental Sciences, KIIT Deemed to be University, Bhubaneswar. Email Id – drmalvikaraghuvanshi@gmail.com

Correspondence:

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Neurofibromatosis Type 2 **[2,3]**. In both sporadic and syndrome-associated cases, neurofibroma development is linked to alterations involving the NF1 gene **[5]**. In sporadic lesions, NF1 mutations are confined to the tumor cells themselves, whereas in syndrome-associated cases, neurofibromas arise as a consequence of a germline mutation in the NF1 gene **[5,6]**. The NF1 gene encodes neurofibromin, a tumor suppressor protein located on chromosome 17q11.2, which plays a critical role in regulating cell growth and proliferation through the RAS signaling pathway **[5,6]**.

Case Presentation

A 19-year-old female presented with a painless swelling over the left forehead for four years. The swelling has an insidious onset with gradual enlargement; no preceding trauma, there was no complaints of headache, fever or recent weight loss, visual disturbance, or focal neurological symptoms. Personal or family history of NF1 or other genetic disorders was nil. On local examination a single, well-defined subcutaneous mass over the left frontal region measuring approximately 3 X 2 cm was observed (Figure 1). The lesion was firm, slightly mobile over underlying bone, non-pulsatile, non-compressible, and non-tender. Overlying skin was normal without discoloration, ulceration, or visible vascularity. Systemic and dermatological examination showed no café-au-lait macules, axillary/inguinal freckling or other cutaneous neurofibromas. Neurological examination was unremarkable.

Diagnostic assessment was done with differential diagnosis of Lipoma, epidermoid cyst, fibrous histiocytoma and vascular malformation. An Excisional biopsy under local/general anesthesia with an elliptical incision placed along a forehead skin crease to optimize cosmesis (Figure 2a & b). The lesion was dissected from surrounding subcutaneous tissue and removed en bloc with preservation of adjacent neurovascular structures. Hemostasis achieved and layered closure performed.

Macroscopically, a well-circumscribed, soft, tan-white nodular specimen measuring 3.0 X 1.8 X 1.5 cm was received (Figure 3). Histopathological examination revealed unencapsulated lesion composed of spindle-shaped cells with elongated, wavy nuclei embedded in a collagenous myxoid stroma, lot of scattered mast cells. There was no significant mitotic activity or necrosis (Figure 4a & 4b).

Postoperative recovery was uneventful, the wound healed by primary intention. No sensory deficit or motor impairment. Follow-up at 2 weeks and 12 months showed no evidence of recurrence and satisfactory cosmetic outcome with no new cutaneous lesions on serial skin exams.

DISCUSSION

Neurofibromas are the most common tumor of the peripheral nerve sheath, affecting men and women equally, without racial or ethnic predilection **[7]**. Age of onset is highly variable; however, localized lesions most commonly occur in adults aged 20 to 40 years **[7,8]**. Neurofibromas are of three types namely localized (most common), diffuse, and plexiform **[8]**. While most neurofibromas arise sporadically and carry an extremely low risk of malignant transformation, the plexiform variant is considered pathognomonic for NF1 and is associated with a higher likelihood of malignant change **[8,9]**. Both diffuse and plexiform neurofibromas are more commonly seen in children, with plexiform lesions rarely developing after the age of five **[4,8]**.

Neurofibromas occur across all age groups but are most commonly diagnosed in adolescents and young adults **[7,9]**. Solitary sporadic neurofibromas are not uncommon overall but are less frequently reported in the forehead region compared to the trunk and extremities **[10]**. Because forehead masses are frequently attributed to more common entities (lipoma, epidermoid/dermoid cyst, osteoma, or fibrous histiocytoma), neurofibromas may be overlooked **[10,11]**. The rarity of forehead localization and the broad differential diagnosis for forehead masses can delay definitive diagnosis and treatment **[10,11]**. Pathogenesis of NF1-associated neurofibromas is due to loss of neurofibromin function leading to increased RAS pathway signalling and uncontrolled Schwann cell proliferation **[5,12]**. Solitary sporadic neurofibromas may arise from localized Schwann cell proliferation without identifiable germline NF1 mutations **[12]**. The exact molecular drivers of sporadic lesions are less well characterized but may include somatic NF1 mutations or other pathway alterations **[12,13]**.

Clinically, the forehead masses are commonly benign and differentials include lipomas, epidermoid/dermoid cysts, pilomatricomas, osteomas, and vascular lesions **[11,14]**. Key clinical features that raise suspicion for a neurogenic tumor include a soft-to-firm consistency,

mobility in the subcutaneous plane, and a long-standing, slowly progressive course **[14]**. Tinel's sign (radiating paresthesia on percussion) may be present with nerve-origin tumors but was absent in this case **[14]**. Imaging and histopathology are essential to distinguish neurofibroma from schwannoma (encapsulated, Antoni A/B areas) and from plexiform neurofibroma (diffuse, infiltrative growth often associated with NF1) **[13,15]**.

MRI is the imaging modality of choice for soft tissue nerve sheath tumors **[15]**. Typical features of neurofibroma include a well-defined lesion with homogeneous or heterogeneous T2 hyperintensity and variable enhancement **[15,16]**. The "target sign" (central low signal with peripheral high signal on T2) can be seen in neurofibromas but is not pathognomonic **[16]**. MRI is particularly valuable to assess lesion extent, relationship to adjacent structures, and to exclude intracranial or intraosseous extension in craniofacial lesions **[15,16]**.

Histologically, neurofibromas are unencapsulated and composed of a mixture of cell types within a collagenous or myxoid stroma **[17]**. The presence of spindle cells with wavy nuclei and a background of collagen and mucin is typical **[17]**. Immunohistochemistry shows S-100 positivity (variable intensity) reflecting Schwann cell components **[7,17]**. CD34 may highlight fibroblastic elements **[17]**. Distinguishing features from schwannoma include lack of a true capsule and absence of Antoni A/B architecture **[15,17]**. Malignant peripheral nerve sheath tumor (MPNST) should be suspected if there is high cellularity, nuclear atypia, increased mitotic figures, necrosis, or rapid clinical growth **[18]**.

Treatment

Surgical excision is the mainstay for solitary neurofibromas when symptomatic, enlarging, or cosmetically concerning **[14,18]**. Goals include complete removal with preservation of function and optimal cosmetic outcome **[18]**. In the forehead, incision planning along natural creases and careful soft tissue handling minimize scarring **[18]**.

Conservative management with observation may be appropriate for small, asymptomatic lesions with low suspicion for malignancy **[18]**. In complex cases such as plexiform neurofibromas or lesions in critical anatomical locations may require staged resections, multidisciplinary planning (neurosurgery, plastic surgery, ENT, ophthalmology), or adjunctive therapies **[18,19]**.

Adjuvant therapies like radiotherapy or chemotherapy have shown limited role in benign neurofibromas; targeted therapies (e.g., MEK inhibitors) have shown benefit in symptomatic, inoperable plexiform neurofibromas associated with NF1 but are not standard for solitary sporadic lesions **[19]**.

Surgical considerations include identification and preservation of sensory nerves, avoidance of injury to the frontal branch of the facial nerve, and meticulous hemostasis **[18]**. Potential complications include sensory disturbance, hematoma, infection, wound dehiscence, and unsatisfactory scar/cosmetic outcome **[18]**. Recurrence after complete excision of solitary neurofibroma is uncommon but possible if excision is incomplete **[18]**.

Prognosis for solitary neurofibroma after complete excision is excellent **[18]**. Malignant transformation is rare in isolated lesions but remains a concern in NF1-associated plexiform neurofibromas affecting approximately 10% of patients with NF1 **[8,9]**. Recommended follow-up includes periodic clinical examination to detect recurrence or new lesions; imaging is reserved for suspicious clinical changes **[18,19]**. In young patients, counselling regarding signs of NF1 and periodic dermatologic and ophthalmologic screening is prudent when clinical suspicion exists **[19]**.

CONCLUSION

Solitary neurofibroma of the forehead is an uncommon but important diagnostic consideration for long-standing forehead swellings. The case emphasizes the importance of considering nerve sheath tumors in the differential diagnosis of chronic forehead masses. Facial lesions can have significant psychosocial effects, particularly in adolescents and young adults. Addressing cosmetic concerns, providing preoperative counselling about expected outcomes and scars, and offering psychological support when needed are important components of holistic care. Thorough clinical assessment, appropriate imaging, and histopathological confirmation guide management. Surgical excision with careful technique yields excellent functional and cosmetic results. Long-term clinical follow-up is recommended to monitor for recurrence and to evaluate for any features suggestive of syndromic disease.

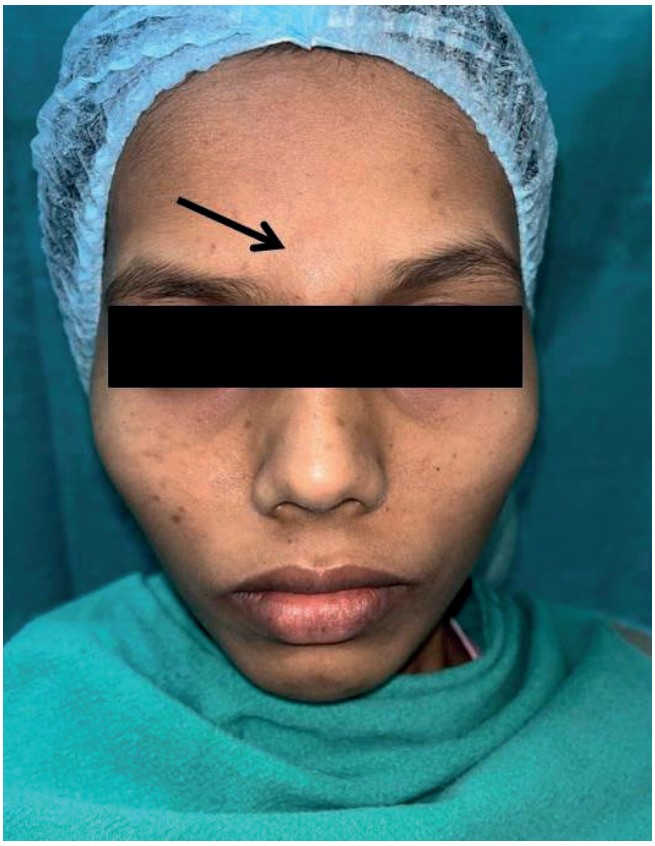

Figure 1: Well-defined subcutaneous mass over the left frontal region

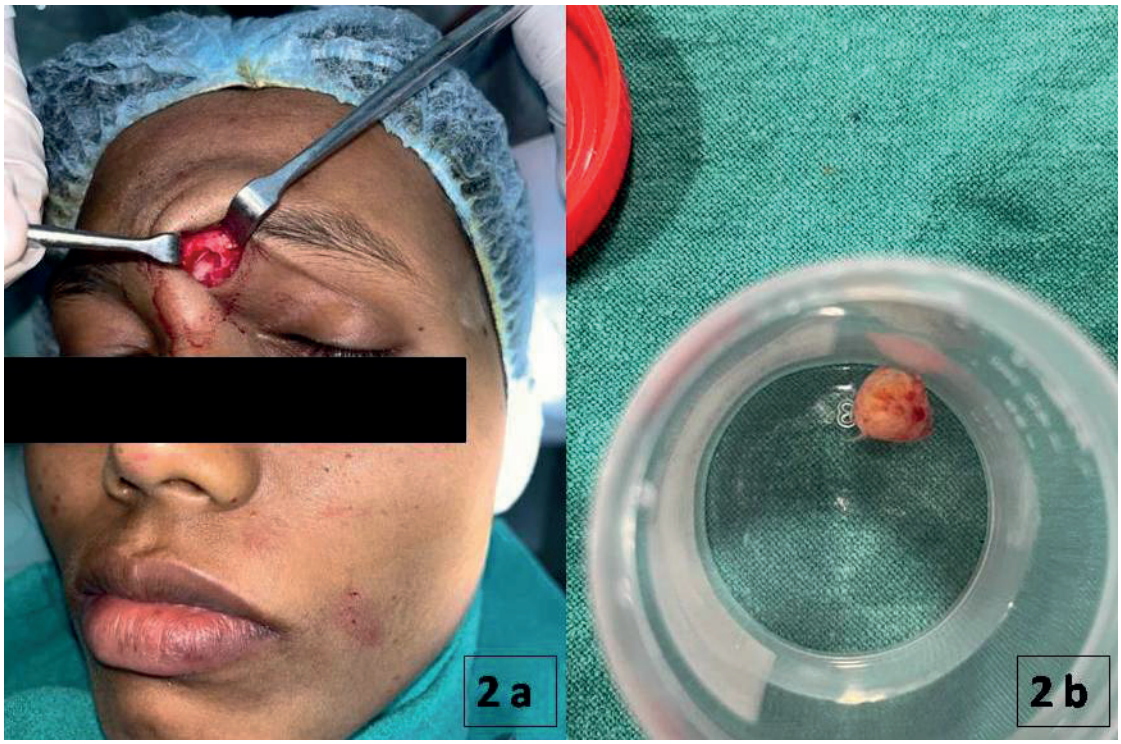

Figure 2a: Surgical excision of the mass **Figure 2b:** Excisional biopsy specimen collected in 10% formalin

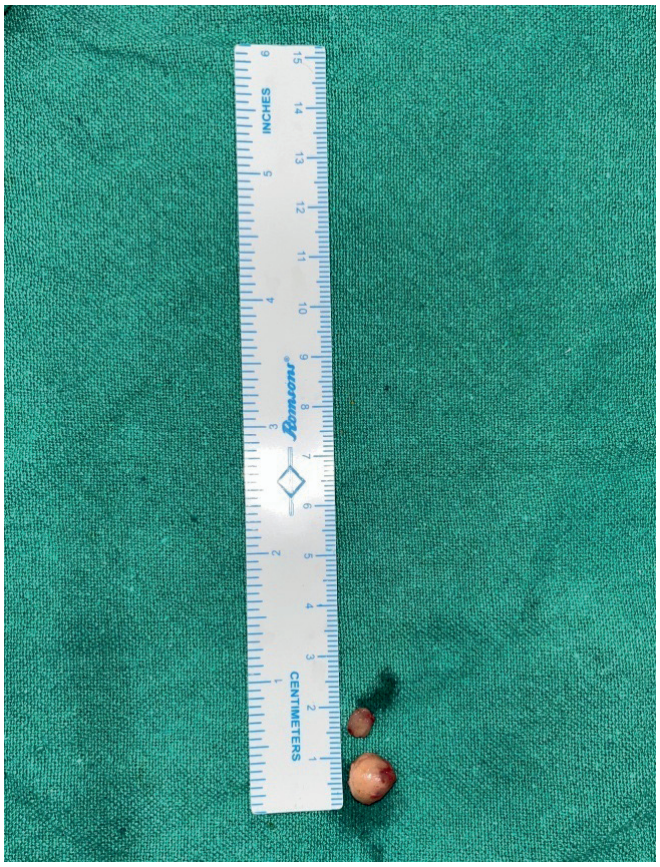

Figure 3: Grossing of the mass received measuring 3.0 X 1.8 X 1.5 cm

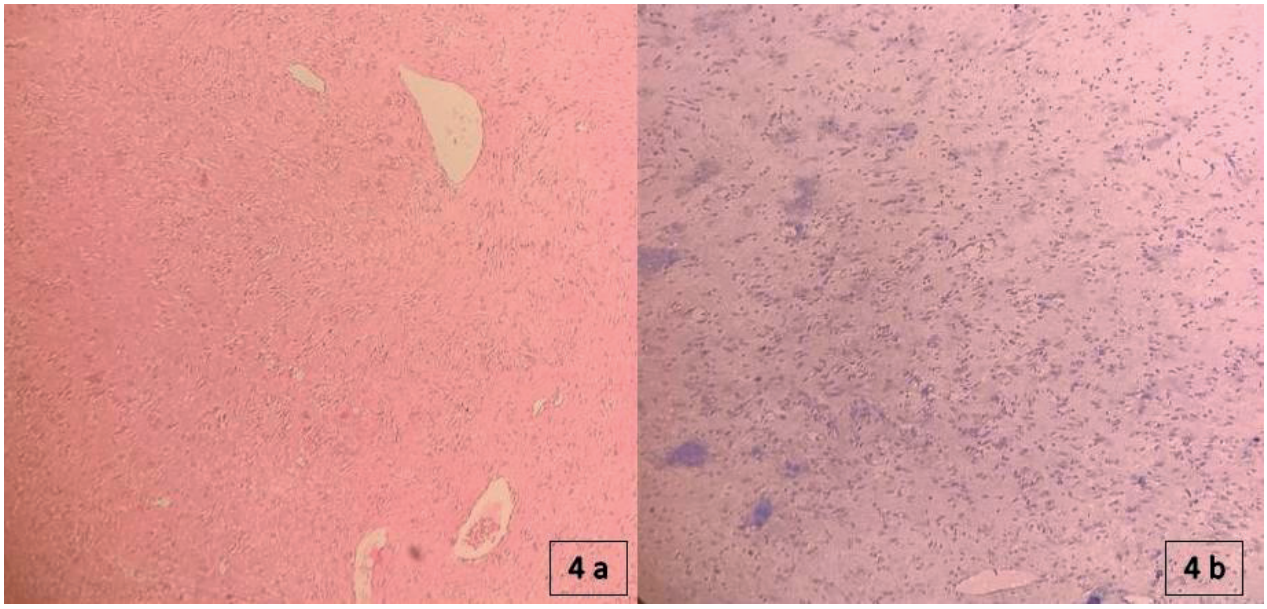

Figure 4a: 4 X magnification shows spindle-shaped cells wavy nuclei

Figure 4b: 10X magnification shows elongated,

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embedded in a collagenous myxoid stroma