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Case Report

Diffuse neurofibroma of the scalp: a distinct histopathological entity

Sreemani Kumari Neelam[1], Padma Malini Kyatham[1], Vijaya Nirmala Byrapuram[1], Lakshmi Narasimha Moorthy Nimmagadda[2], Shravan Kumar Odapally[1]

[1]Department of Pathology, Gandhi Medical College - Dr.NTRUHS, Hyderabad, Telangana, India
[2]Department of Radiology, Gandhi Medical College - Dr.NTRUHS, Hyderabad, Telangana, India

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***Correspondence:**
Dr. Sreemani Kumari Neelam,
E-mail: nsmk13959@gmail.com

ABSTRACT

Neurofibroma is a benign tumour of peripheral nerve sheath characterized by proliferation of Schwann cells, perineurial cells and endoneurial fibroblasts, involving the skin and the subcutaneous tissue in a plaque like fashion on the head and neck regions, primarily in adolescents and young adults. Different types of Neurofibromas can be identified including localized, plexiform and diffuse types. The diffuse variant is rare and distinct form, both histologically and radiologically. Excision biopsy, histopathological examination is the main diagnostic tool. S100 protein was positive. We present a case of recurrent Diffuse Neurofibroma over the Scalp in a 20 year old male patient, in the background of Neurofibromatosis 1.

Keywords: Peripheral nerve sheath tumours, Diffuse neurofibroma, Von Recklinghausen's disease, S100 Protein

INTRODUCTION

Diffuse Neurofibroma is an uncommon, but distinct variant of Neurofibroma affecting the trunk, head and neck regions of adolescents and young adults. It is not clearly known how diffuse Neurofibromas are associated with Neurofibromatosis, although it has been suggested that 10% of cases of diffuse Neurofibromas have Von Recklinghausen's disease NF-1.[1,2] We report a case of recurrent Diffuse Neurofibroma of the scalp in a 20 year old young adult male patient, which was confirmed by radiological imaging and characteristic histopathogical findings.

CASE REPORT

A 20 year old male patient was admitted under surgical speciality for recurrent scalp swelling. The initial age at presentation was when he was 2 years old. Surgery was attempted and tumour was removed when the patient was 10 years old. At present the patient came back with a history of recurrent scalp swelling, gradually increasing in size and has attained the present size since 2 years, i.e. when the patient was 18 years of age.

Physical examination revealed a 20 x15 cms, oval to oblong enplaque swelling over the scalp, soft in consistency, non-cystic, non-tender with no visible pulsations or bruit on auscultation over the swelling. The swelling was seen occupying left parieto-temporal regions of the scalp and extending on to the opposite side crossing the midline. The overlying skin was normal excepting for the old scar on the left lateral side of the scalp.

Radiographs of the skull did not reveal any bone erosion. CT scan of the head showed an extra cranial soft tissue mass on the left side of the scalp extending on to the right side with no intracranial extension. The underlying bone showed focal thickening. MRI of the brain with contrast

on T1-weighted imaging revealed linear or reticular strands of intermediate signal intensity in the subcutaneous fat, indicating the tumour tissue along with collagen and ecstatic vessels, while on T2-weighted imaging were of high signal intensity, in contrast to collagen fibres.[3]

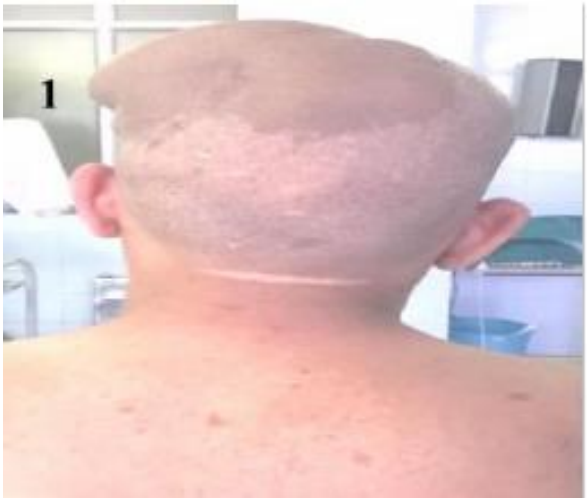

Figure 1: Posterior views of clinical photographs showing the left parietal enplaque like scalp mass.

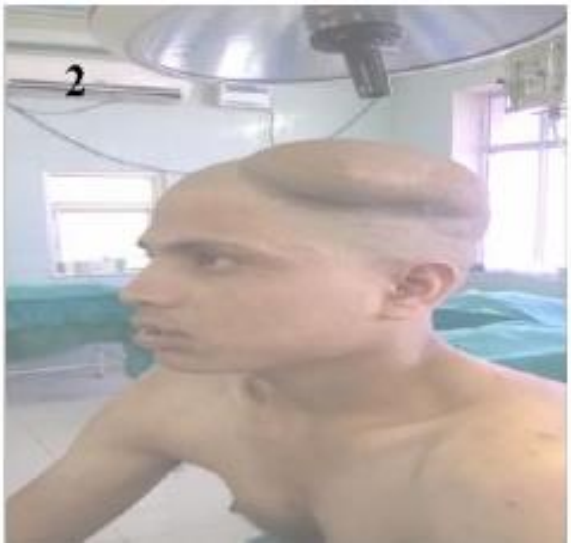

Figure 2: Lateral views of clinical photographs showing the left parietal enplaque like scalp mass.

Fine needle aspiration cytology was done which revealed few mature adipocytes along with blood elements. A diagnosis of Lipoma was made. Excision biopsy was done with skin grafting and the specimen was submitted for histopathological examination.

Pathology

Received multiple grey white to grey brown soft tissue masses, largest and smallest masses measuring 11 x 8 x 1 cm and 1.5 x 1.5 x 1 cm in dimensions respectively. Cut section of the masses revealed grey white, glistening, homogenous appearance with spotty areas of haemorrhages. Multiple H & E stained sections studied revealed a tumour tissue arranged in whorls, fascicles admixed with proliferated blood vessels. Cells were fusiform, with elongated nuclei and surrounded by a uniform myxoid matrix of wire-like fine fibrillary collagen fibres and admixed with plenty of Wagner-Meissner's bodies.[4] There was no evidence of any pleomorphism, necrosis, increased mitoses or bizarre tumour giant cells. A diagnosis of diffuse neurofibroma was made. IHC studies with S100 protein were strongly and diffusely positive, thus complementing the diagnosis.[5] Thorough examination of the patient for stigmata of Neurofibromatosis 1, revealed cafe-au-lait spots all over the body, especially over the nape of the neck, back and shoulder regions ranging in size from 0.3 to 1.5 cm in diameter. No lisch nodules were identified.

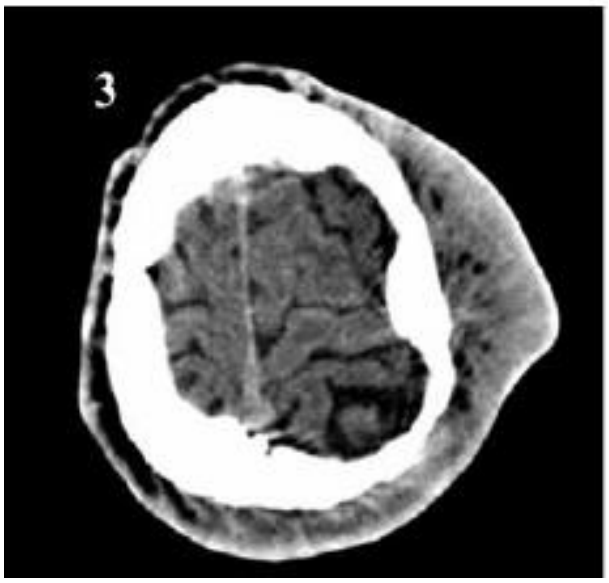

Figure 3: CT – Brain: shows diffuse extra cranial soft tissue density mass with minimal calvarial thickening.

The patient's physical and mental development revealed no abnormalities. Follow up of the patient was uneventful. A final diagnosis of Recurrent Diffuse Neurofibroma of the scalp in the background of Neurofibromatosis-1 was made.

DISCUSSION

Neurofibromas are benign tumours of the peripheral nerves of neuroectodermal origin. The essential cells in neurofibroma are of Schwann cell origin. Diffuse neurofibroma is an uncommon, but distinct form of neurofibroma, both clinically and histologically. It is variable in size, though often large. It is characterized by marked dermal and subcutaneous thickening that most often appears in the trunk or head & neck regions of young adults. Intracranial extension of the tumour has rarely been reported.[6] This lesion has also been termed as

paraneurofibroma indicating its extension beyond the confines of the perineurium.[7] At least 10% of tumours are associated with NF-1, although Megahed reported that 61% of his 13 patients with diffuse neurofibromas had NF-1.[7]

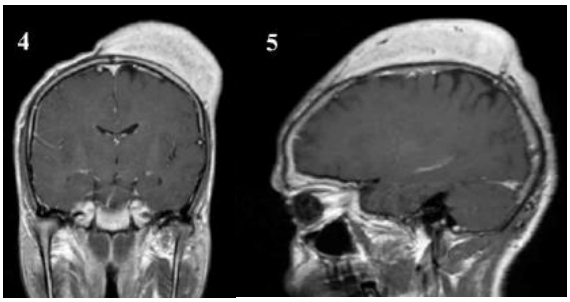

Figure 4 and 5: MRI with contrast: Show diffuse enhancing purely extra cranial mass along the left parietal convexity. Intracranial structures are normal.

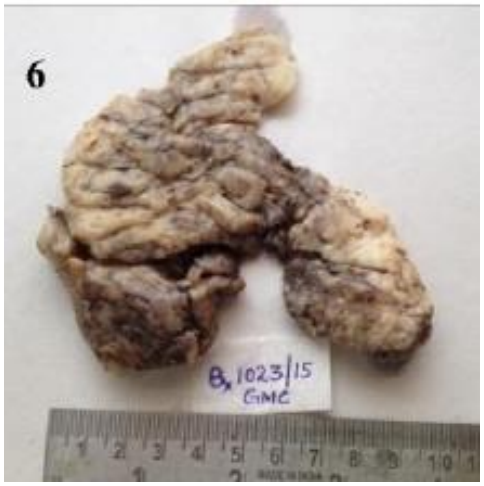

Figure 6: Gross photographs of the scalp tumour.

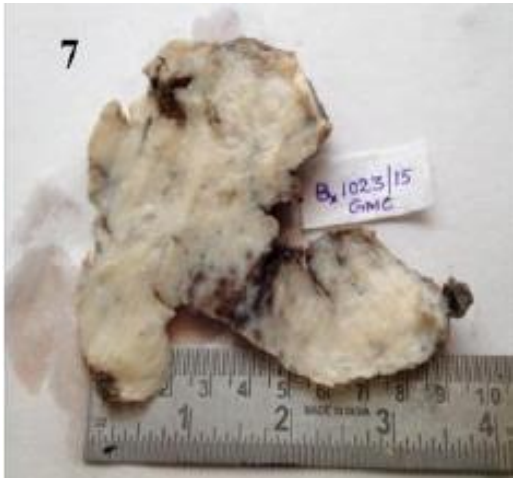

Figure 7: Cut section: shows grey white scalp tumour.

The imaging procedures to diagnose Diffuse Neurofibromas include, Ultrasonography, CT-Scan & MRI. MRI is the investigative tool of choice for the diagnosis, because it better defines the anatomic relationship between neurofibroma and adjacent tissues, like muscles, vessels and neural structures. MRI differentiates diffuse neurofibroma from those of the localized and plexiform types of neurofibroma. On MRI, diffuse neurofibromas exhibit typical target pattern, revealing plaque-like infiltrating masses involving the skin and subcutaneous tissue characterized by the presence of linear or reticular strands. These strands are of hypointense signal intensity on T_1 weighted images and appear hyperintense on T_2 weighted images in comparison with muscle. Localized neurofibroma appears as a well-defined nodule while plexiform neurofibroma appears as a serpiginous lesion. The MRI findings of our patient correlated with all the above findings.

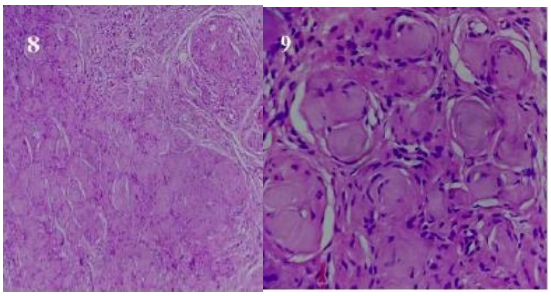

Figure 8 and 9: H&E: 10x & 40x Views: Shows tumour tissue arranged in whorls, fascicles composed of fusiform cells with elongated nuclei and surrounded by a uniform myxoid matrix of wire-like fine fibrillary collagen fibres.

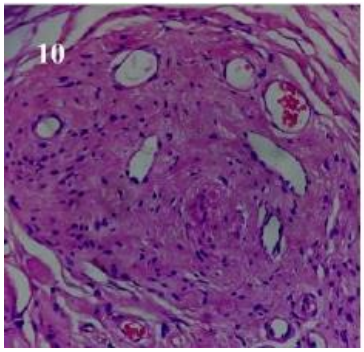

Figure 10: H&E: 40x View: Shows wagner-meissner's body.

On USG & CT[8], Diffuse Neurofibroma may resemble an intramuscular haemangioma or Lipoma due to the increased vascularity & presence of adipocytes intermingling with neurofibroma cells & also due to entrapment of perineural adipose tissue by diffuse neurofibromas.

On histopathology the tumour is ill defined, diffusely infiltrating the dermis and subcutaneous tissues. Despite its infiltrative nature, the tumour envelops the normal structures rather than infiltrate or destroy them. Cells are

fusiform, with elongated nuclei and are surrounded by a myxoid matrix of wire-like collagen fibres. Wagner Meissner's bodies are characteristic, but may not be always present. Neurofibromas show immunoreactivity with S100 protein, a sensitive but not a specific marker for benign nerve sheath tumours. Although malignant transformation of neurofibromas has been reported in patients with NF1 it rarely occurs in diffuse neurofibromas. Pain or enlargement of a neurofibroma may herald the malignant transformation.

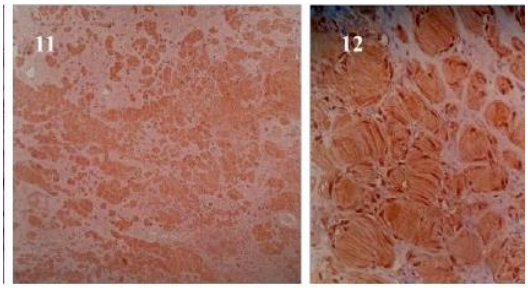

Figure 11 and 12: 10x & 40x Views – IHC: Diffuse & intense positivity of S-100 protein.

Megahed described 10 histopathological variants of neurofibroma: classic, cellular, myxoid, hyalinised, epithelioid, plexiform, diffuse, pigmented, granular cell, and pacinian. Subsequently, some other variants such as dendritic cell neurofibroma with pseudo rosettes and lipomatous neurofibroma have been reported. The treatment of large neurofibromas consists of partial or complete excision, especially for isolated cases. Because of the rich vascular supply, pre-operative angiogram and intra-arterial embolization is helpful in reducing the risk of haemorrhage. Even after complete excision, clinical recurrences may develop because of the infiltrative growth pattern of the tumour. Because of possible recurrence and potential development of neurofibromatosis, yearly follow up of the cases is recommended. In the present case the imaging studies and clinical findings demonstrated an infiltrative pattern of growth. Consecutive surgeries were done for complete excision.

In summary, we report a case of recurrent Diffuse Neurofibroma of the scalp in a young adult in the background of NF-1. This is a rare peripheral nerve sheath tumour with characteristic radiological appearances that correlates well with the distinct histopathology. The reticular appearance is due to a network of abnormal nerves set in a collagen background. It is known to be demonstrated accurately on MRI, particularly contrast enhanced T1-W & T2-W sequences. However confirmation is by the characteristic histopathology in conjunction with IHC correlation. The post-operative period of our case was uneventful and regular follow-up was advised.

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REFERENCES

1. Yoo AKH, Kim BJ, Rho YK, Lee JW, Kim YK, Kim MN, et al. A Case of Diffuse Neurofibroma of the Scalp. Ann of Dermatol. 2009;21(1):46-8.
2. Kumar BCS, Gopal MG, Talwar A, Ramesh M. Diffuse Neurofibroma of the Scalp Presenting as Circumscribed Alopecic Patch. Int J Trichology. 2010;2(1):60-2.
3. Huang GS, Huang CW, Lee HS, Chang WC, Lee CH, Leu NH, et al. Diffuse Neurofibroma of the Arm: MR Characteristics. 2005;184(5).
4. Cha SH, Cho SH, Lee JD. A Case of Diffuse Neurofibroma. Korean J Dermatol. 2008;46(3):355-38.
5. Zuuren EJV, Posma AN, et al. Diffuse neurofibroma on the lower back. J Am Acad Dermatol. 2003;48(6):938-40.
6. Kumar S, Bhaskar S, Handa A, Jindal B. Diffuse neurofibroma of scalp Asian J Neurosurg. 2014;9(4):237.
7. Khan AK, Deb S, Ray dk, Mandal S, Mukhopadhaya S, Mandal S. Diffuse neurofibroma of scalp; Neurology India. 2002;50(4):516-7.
8. Hassell DS, Bancroft LW, Kransdorf MJ, Peterson JJ, Berquist TH, Murphey MD, et al. AJR Am J Roentgenol. 2008;190(3):582-8.

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