Case Report

Neurofibroma with Bone Destruction Masquerading Extracranial Meningioma

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Abstract

Neurofibromatosis 1 (NF1) an autosomal dominant disorder, is the most common of the phakomatoses (neurocutaneous syndromes) and has a variety of localized or systemic manifestations and the classic systemic lesions include neurofibromas and more specifically, plexiform neurofibromas. A 60-year-old female presented with a progressively increasing swelling over left side of forehead and right lumbar region associated with pain. In addition she had multiple painless swellings all over body. Imaging findings of the brain were suggestive of extracranial meningioma in present case; however the histopathology confirmed the diagnosis of neurofibroma.

Key words: Neurofibroma, Extracranial Meningioma, Meningioma

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Introduction

NF1 an autosomal dominant disorder, is the most common of the phakomatoses (neurocutaneous syndromes) that occurs in one of every 2,000 live births. NF1 has a variety of localized or systemic manifestations and the classic systemic lesions include neurofibromas and more specifically, plexiform neurofibromas [1-5].

Case Report

A 60-year-old female presented with a progressively increasing swelling over left side of forehead and right lumbar region associated with pain. In addition she had multiple painless swellings all over body. Her general and systemic examination was normal. Higher mental functions were normal. There were no focal neurological deficits. Local examination revealed a large painful swelling over

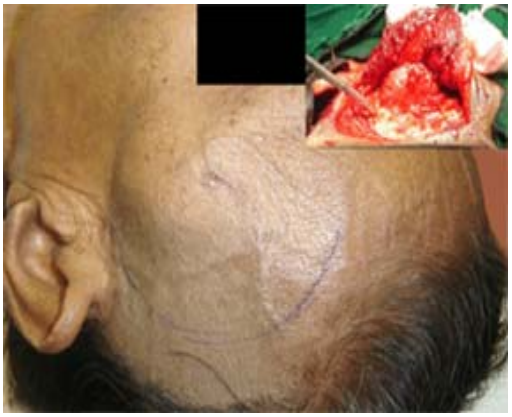

Figure 1. *Clinical photograph showing swelling over left side of forehead*

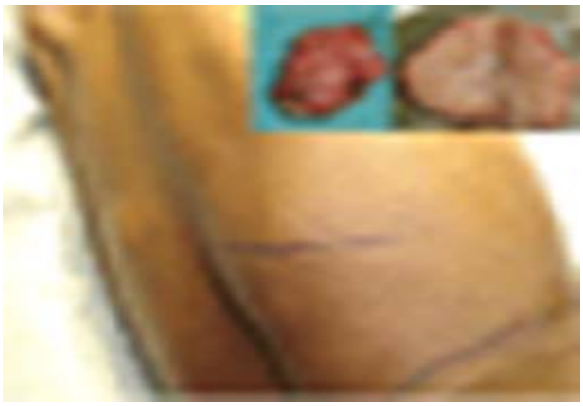

Figure 2. *Clinical photograph showing swelling over right lumbar region*

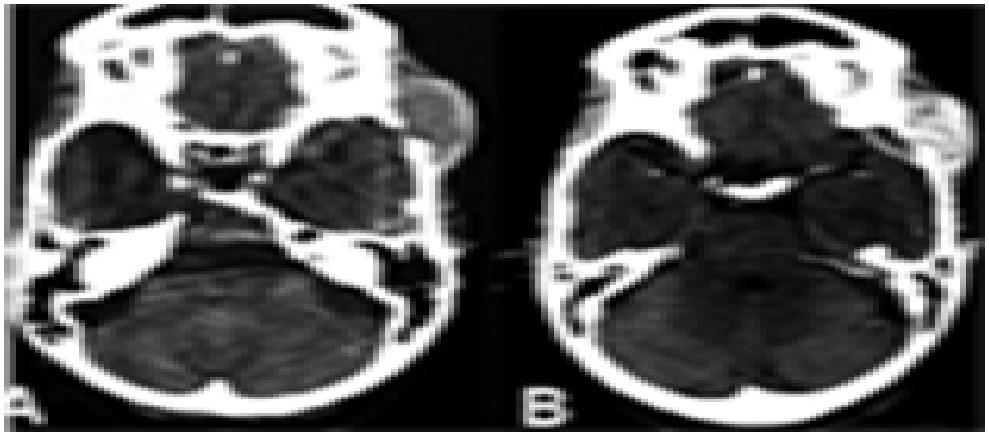

Figure 3. *Computed tomography scan showing the tumor with homogenous enhancement and erosion of the adjoining bone is present*

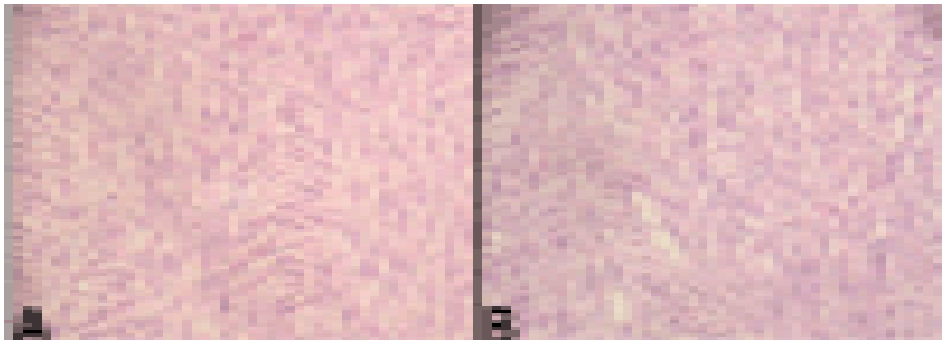

Figure 4 (A and B). *Histopathology from both the lesions was suggestive of Neurofibroma*

left side of the forehead and a large swelling over right lumbar region (Fig 1 and 2). Computed tomography (CT scan) of the brain showed a hyperdense, enhancing mass involving left pterional region with the destruction of bone and associated with hyperostosis of the bone (Fig 3). Based on the clinical features and imaging findings a diagnosis of left pterional extracranial meningioma with multiple neurofibroma was made. The patient underwent surgical of the both the lesions. On histopathological examination of both the lesions there was presence of spindle shaped cells with wavy and tapered nuclei arranged in fascicles, features suggestive of neurofibroma (Fig 4).

Discussion

The typical clinical picture of NF1 includes spots of hyperpigmentation (cafe´ au lait spots) and cutaneous and subcutaneous tumors [4]. In addition there may be axillary freckling, iris hamartomas, bone dysplasias, affected first degree relatives, and multiple central nervous system tumors [6]. Long standing neurofibroma can lead to the bony destruction and confirmation is possible only after biopsy [7-9]. In present case the lesion was confused with the extracranial meningioma as the appearance was mimicking the meningioma (bone destruction and attachment to dura) [10].The mainstay of treatment for cutaneous neurofibromas is surgical removal; however carbon dioxide laser may be indicated for small, superficial lesions [1-5].

Acknowledgment

None.

Conflicts of Interest

The authors have no conflicts of interest in this article.

Author's Contribution

All the Authors have equally contributed in the preparation of manuscript, literature review and search.

References

1. Fortman BJ, Kuszyk BS, Urban BA, Fishman EK. Neurofibromatosis Type 1: A Diagnostic Mimicker at CT. RadioGraphics 2001; 21:601–12.

2. Kumar AJ, Kuhajda FP, Martinez CR, Fishman EK, Jezic DV, Siegelman SS. Computed tomography of extracranial nerve sheath tumors with pathological correlation. J Comput Assist Tomogr 1983; 7:857–65.

3. Reed JC, Hallet KK, Feigin DS. Neural tumors of the thorax: subject review from the AFIP. Radiology 1978; 126:9–17.

4. Biondetti PR, Vigo M, Flore D, De Faveri D, Ravasini R, Benedetti L. CT appearance of generalized von Recklinghausen neurofi- bromatosis. J Comput Assist Tomogr 1983; 7: 866–9.

5. Friedman JM, Gutmann DH, MacCollin M, Riccardi VM: Neurofibromatosis: Phenotype, Natural History and Pathogenesis. Baltimore: Johns Hopkins University Press; 1999.

6. Williams M, Verity CM. Optic nerve gliomas in children with neurofibromatosis. Lancet 1987; 1:1318–19.

7. Hvorslev V, Reiter S. A case of neurofibromatosis with severe osseous disease of the thoracic spine. Pediatr Radiol. 1979; 8(4):251-3.

8. Kaufman A, Sherman FC, Black W, Duban S. Osseous destruction by neurofibroma diagnosed in infancy as "desmoplastic fibroma". J Pediatr Orthop 1984; 4(2):239-42.

9. Baehnisch G, Brückner L. Destruction of the cervical spinal column in neurofibromatosis. (Recklinhausen's disease). Beitr Orthop Traumatol. 1978; 25(9):515-8.

10. Agrawal A, Rao KS, Makannavar JH, Shetty L, Patel N. Extracranial meningioma in the vicinity of the temporal bone: a difficult preoperative diagnosis. Surg Neurol. 2007; 67(1):102-5.