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Case Report

A rare case report on solitary pigmented neurofibroma over scalp region

Patel Pratik Girishbhai*, Lav R. Mehta, Anuj Awasthi, Manoj Talreja, Chetan Prajapati

Department of General Surgery, Dr. M K Shah Medical College and Research Centre and Smt. SMS Multispeciality Hospital, Gujarat University, Ahmedabad, Gujarat, India

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***Correspondence:**
Dr. Patel Pratik Girishbhai,
E-mail: pratikp489@gmail.com

ABSTRACT

Neurofibromas are benign proliferations that are made up of all nerve elements, and arise as fleshy and nontender, sessile or pedunculated masses on the skin. Pigmented (or melanocytic) neurofibroma constitutes only 1% of cases and is considered a rare variant of neurofibroma containing melanin-producing cells. Our patient has solitary pigmented neurofibroma over scalp region which is very rare and only few cases were reported. Here we present a case of 25 year old female patient who had brown colored swelling since birth over left temporo-parietal region which gradually increased in size with age. Patient had no any other swelling or pigmentation over any other regions of body. Patient underwent excision for the same with skin grafting. Her recovery was uneventful.

Keywords: Solitary, Pigmented neurofibroma, Scalp region

INTRODUCTION

Neurofibromas consist of both nerve sheath and nerve cells and account for up to 25% of nerve sheath tumours. Up to 40% of patients with mediastinal fibromas have generalized neurofibromatosis (von Recklinghausen's disease). About 70% of neurofibromas are benign, but malignant degeneration to neurofibrosarcoma occurs in 25% to 30% of patients.[1] It can arise sporadically or in association with type 1 neurofibromatosis often associated with café-au-lait spots and lisch nodules.[1] Neurofibromatosis type 1 (NF1) is an autosomal dominant condition caused by mutation of NF1 gene located at chromosome 17q11.[2]

Neurofibromatosis type 2 (NF2) is associated with meningioma and acoustic neuroma and caused by mutation of NF2 gene located at chromosome 22q12. Pigmented neurofibroma is a rare subtype of neurofibroma, it is considered a chronically progressive benign tumor containing melanin-producing cells. These lesions can appear alone or in association with neurofibromatosis.[3]

A neurofibroma not associated with NF1 is termed solitary neurofibroma. Neurofibromas are known to occur more frequently in the head and trunk than in other parts of the body.[4] In 90% of cases, neurofibroma present as solitary lesions, with the remainder presenting in patients with NF1, an autosomal dominant genetically inherited disease. NF1 occurs in 1 in 3000 births and has been referred to as peripheral neurofibromatosis or von Recklinghausen disease.[5]

NF2 is a genetically determined disorder which affects 1 in 40000 individuals worldwide. A diagnosis of NF2 is made when an individual has the following findings: schwannomas on both 8th cranial (vestibular) nerves or a parent, sibling or child with NF2 plus one vestibular schwannoma in a person less than 30 years of age, or any two of the following: meningioma, glioma, schwannoma, juvenile cataracts.[5]

CASE REPORT

Presenting a case of 25 year old female patient who had brown colored swelling over left temporo-parietal and occipital region of scalp. It was present since birth which gradually increased in size till the present size of approx. 13×13 cm. It was associated with itching and not associated with headache, redness, tenderness, weight loss, tingling or numbness. There is no associated family history or previous surgical history.

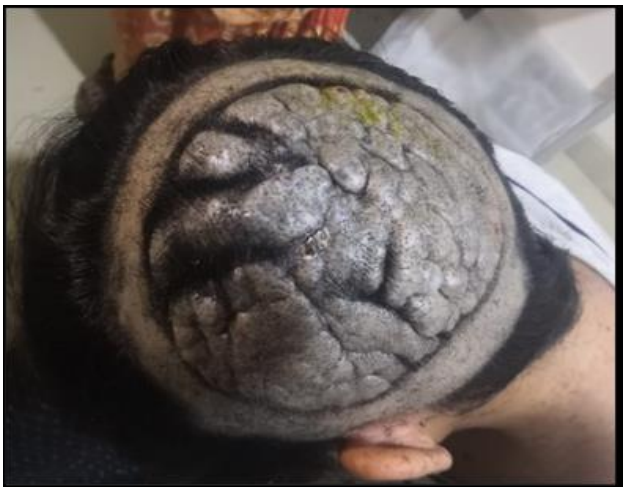

Figure 1: Preoperative findings of swelling.

On examination

Patient is conscious, cooperative and oriented to time, place and person. Patients has normal BMI and her vitals are within normal range. Patient has no any hearing abnormality or any neurological deficit.

A single diffuse swelling of size 12.5×12.5×1.5 cm^3 which was firm in consistency with irregular and well-defined margins and had cerebriform like surface containing hairs was present, involving left temporo-parietal region, occipital region and a part of right parietal region. Skin over the swelling is dry with no rise in temperature, redness, tenderness or any pus discharge. There was fixity to skin with slip sign, fluctuation test and transillumination test were negative. Swelling was non-reducible, non-compressible and non-pulsatile in nature. There was no any other pigmentation or swelling present over other parts of body.

Investigations

All the routine blood investigations like CBC, RFT, LFT, electrolytes were in normal range. Patient was non-reactive for HBsAg, HIV and HCV.

USG of affected part suggested cerebriform epidermal naevus is more likely over multiple Trichilemmal cysts. MRI brain was done which showed no intracranial extension and intracranial abnormality. Punch biopsy from the affected part was done which suggested benign neural sheath tumor-neurofibroma.

Surgical technique

All the preoperative preparations like shaving of local part and preparation of the donor site for skin graft was done one day before surgery. Prophylactic antibiotic given prior to surgery.

Under general anesthesia, prone position given. Using sterile marker skin was marked 5mm away from the lesion. Dissection started by placing skin incision over left side of the lesion, and deepened dissecting connective tissue, epicranial aponeurosis and loose areolar tissue to reach pericranium.

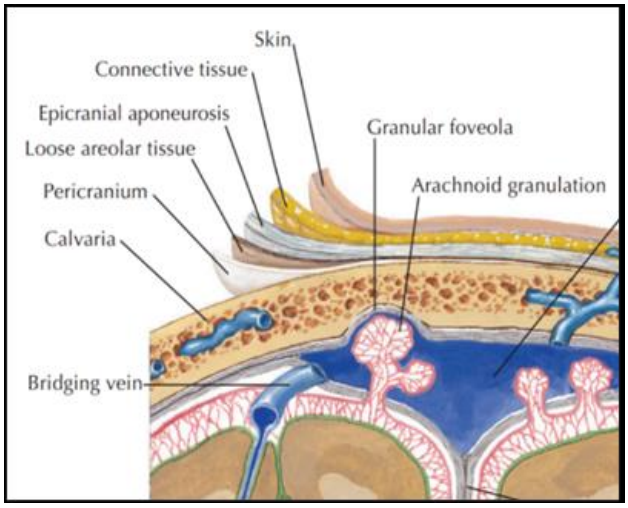

Figure 2: The layers of the scalp.[6]

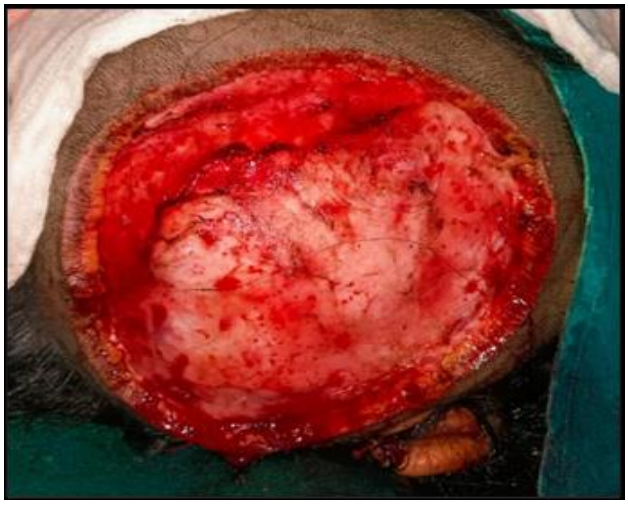

Figure 3: Intraoperative finding after removal of lesion.

Dissection continued towards the opposite side along the plane between loose areolar tissue and pericranium, lesion was excised and sent for histo-pathological

examination. Hemostasis achieved. Under all aseptic precautions, Split thickness skin graft was taken from left thigh and kept over the recipient area. Tie over- Bolster dressing kept. Procedure was uneventful.

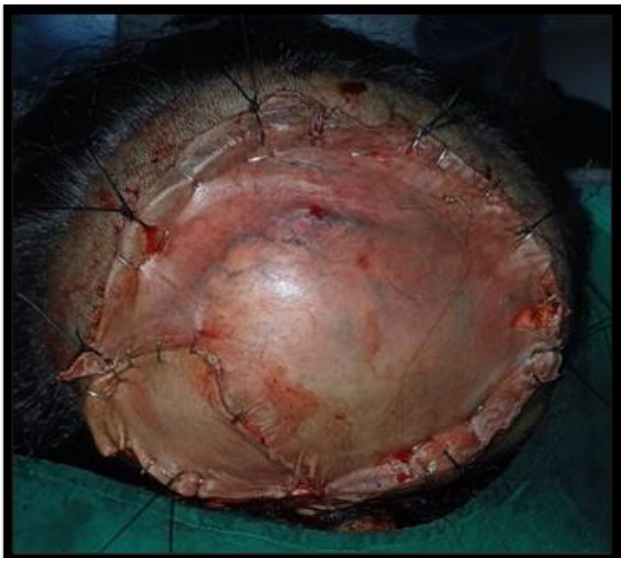

Figure 4: Intraoperative finding after placing skin graft.

After 7 days the dressing was opened, the graft was well taken up with no blackening or discharge.

On histo-pathological examination

On gross examination, the specimen consists of single skin covered flap like soft tissue structure having whitish inner surface and greyish and nodular outer surface with presence of hair.

On microscopic examination, the section shows atrophic dermatitis. Upper dermis is normal. Deep dermis and subcutaneous tissue show presence of benign spindle cells and epitheloid cells arranged in fascicles and whorled pattern. There are foci of melanin pigmentation in spindle cells. No evidence of malignancy is seen. Above all findings suggestive of pigmented neurofibroma.

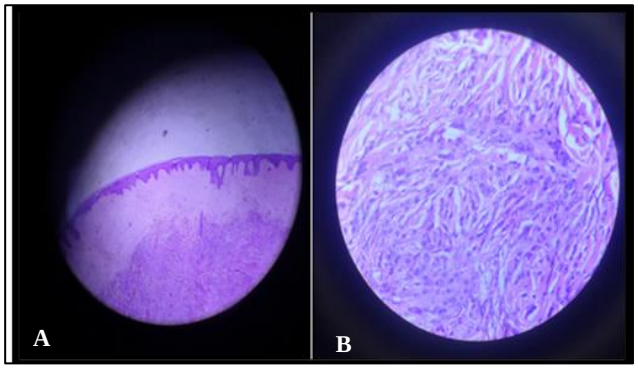

Figure 5 (A and B): Histo-pathological findings.

DISCUSSION

NF1 was first described by the German pathologist Friederich Daniel von Recklinghausen in 1882. It is the most common neurocutaneous syndrome. Diagnosis of NF1 is based on the presence of at least two of the seven criteria established by the national institute of health (NIH) consensus development conference in 1988.[7]

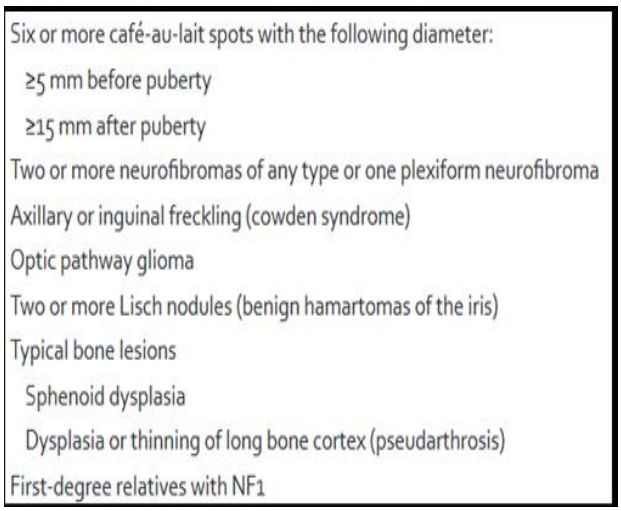

Figure 6: Diagnostic criteria for NF 1 (Two or more of the criteria are required).

Pigmented neurofibroma is a rare variant of neurofibroma that has often been confused with other pigmented cutaneous lesions. Typical histomorphologic features with unique patterns of melanogenesis, histochemical stains and immunohistochemistry enable diagnosis of PNF accurately and also help differentiate it from other pigmented tumours.[8]

Pigmented neurofibroma is defined by the strong pigmentation of cells within the tumour. Melanotic neurofibroma has been described for both NF1 and sporadic manifestations.[9]

Pigmented neurofibromas should be differentiated from the pigmented forms of dermatofibrosarcoma protuberans (Bednar tumour), blue nevi and melanotic schwannoma.[10]

In our case, patient had solitary pigmented neurofibroma over scalp region and there were no other findings suggestive of its association with NF1. The diagnosis of pigmented neurofibroma became possible due to patients' history, clinical examination and histopathological findings.

CONCLUSION

Because of the lack of distinctive clinical characteristics, especially in isolated lesions, it is difficult to diagnose the lesion preoperatively, and such lesions, remain largely, a histopathologic diagnosis. Proper history-taking, physical

examination, opthalmologic and radiodiagnostic investigations are warranted in such cases, with special attention to detection of cafe-au-lait spots, Lisch nodules, and bilateral acoustic to exclude neurofibromatosis.

We reported a case of localised solitary pigmented neurofibroma on the scalp region of a girl which was precent since birth. Patient had no any hearing abnormality or any neurological deficit. The imaging studies and clinical findings didn't demonstrated any infiltrative pattern of growth, and hence complete excision with a skin graft was the best option.

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