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Clinicopathologic Challenge

Unusual Nodular Aggregation in the Retroauricular Area

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CASE DESCRIPTION

A 27-year-old male patient presented with a painless swelling on the right retroauricular area, which has been gradually increasing in size for the past 5 years. There were no signs of itching, pain, discharge, paraesthesia or hearing impairment. History of any preceding trauma or similar findings among family was negative.

The skin examination revealed a collection of soft, painless and skin-coloured nodules in the right retroauricular area with normal overlying skin, ranging from 1 to 3 cm in size [Figure 1a]. Buttonhole sign was positive. On dermoscopic evaluation, a yellowish-brown background with exaggerated skin markings was noted [Figure 1b]. There were no other cutaneous or systemic abnormalities. High resonance computed tomography (CT) showed an extra calvarial soft-tissue swelling of a maximum thickness of 7.5 mm in the right temporal region without any bony invasion or intracranial extension.

HISTOPATHOLOGIC FINDINGS

A punch biopsy was taken for histopathological examination (HPE) which depicted randomly oriented spindle cells with wavy, hyperchromatic nuclei and fine collagen fibres on a background of myxoid stroma without cellular atypia [Figure 2a and b]. Immunohistochemistry was positive for S-100 [Figure 2c].

DIAGNOSIS

A final diagnosis of non-syndromic solitary neurofibroma (SNF) was made and the patient underwent complete surgical excision.

DISCUSSION

Neurofibromas are benign nerve sheath tumours composed of Schwann cells, perineural-like cells, fibroblasts and mastocytes. It represents about 5% of all benign tumours found in soft tissues.[1] In 90% of the cases, these tumours occur as sporadic, solitary non-syndromic tumours with a very low risk of malignancy while the remaining cases are associated with neurofibromatosis type 1 (NF1).[2]

Tumours such as lipoma, benign appendageal tumour and epidermal inclusion cyst were the clinical differential diagnosis considered in our case. They were excluded based on its soft consistency, absence of a central punctum and limited mobility.

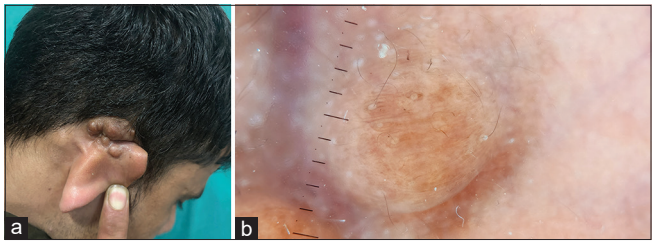

Figure 1: (a) Retroauricular area showing aggregated nodules with positive Buttonhole sign. (b) Dermoscopy shows exaggerated skin markings against a yellowish-brown background (Dermlite DL 5 ×10 magnification, polarised mode).

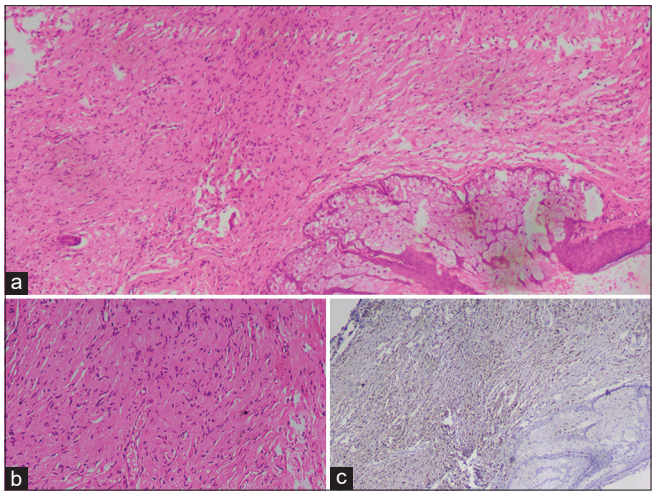

Figure 2: (a) Histopathology shows randomly oriented spindle cells with wavy, hyperchromatic nuclei without any cellular atypia (haematoxylin-eosin; original magnification ×10). (b) Histopathology shows interlacing bundles of elongated cells with wavy darkly stained nuclei (haematoxylin-eosin; original magnification ×40). (c) Immunochemistry showing S100 positivity.

Radiological examination such as ultrasonography, magnetic resonance imaging and CT helps in determining the location and morphology of the tumour. The gold standard for diagnosis is HPE, which reveals similar findings to our case. On immunohistochemistry, S-100 protein, CD-34, cholinesterase activity, vimentin and myelin basic protein are positive markers.[3]

Spindle cell tumours consist of a heterogenous group of cutaneous neoplasms imposing a diagnostic challenge.[4] Tumours commonly encountered include dermatofibroma, dermatofibrosarcoma protuberans (DFSP), plexiform neurofibroma (PNF) and schwannoma. Dermatofibroma appears as firm, non-tender nodules of size up to 1 cm on the extremities. They are tethered to the skin surface with positive Dimple sign. It is histologically characterised by the proliferation of mononuclear, spindle to round or histiocytic cells arranged in a storiform fascicular pattern. Immunohistochemically, the cells are positive for factor XIIIa and vimentin. While, DFSP is a skin sarcoma which appears as an indurated plaque with multiple nodules adherent to underlying structures predominantly on the trunk. Histologically, it exhibits more cellularity with pronounced monotonous, storiform growth pattern which may extend into subcutaneous fat with a characteristic honeycomb appearance. It shows diffuse and strong CD34-positive immunostaining. PNF is a well-circumscribed tumour of the peripheral nerve sheath involving multiple nerve fascicles associated in 5–15% cases of NF-I. They are slow-growing deforming masses giving a 'bag of worms' consistency. On HPE, it displays multiple intertwined hypertrophic nerve fascicles of a subcutaneous or deep nerve embedded in cellular matrix with or without atypia and degenerative changes. While schwannomas are composed of only Schwann cells. They present as a solitary dermal or subcutaneous nodule. On HPE, they are circumscribed, encapsulated by perineurium and are characterised by two types of histological patterns: Antoni type A and Antoni type B. Antoni A is a highly ordered cellular pattern in which spindle cells are arranged in compact fascicles and their nuclei are arranged in palisades. Verocay bodies are a characteristic feature in Antoni type A pattern, with collagen matrix arranged into palisading. Antoni type B tissue exhibits a looser structure of mucinous matrix and it's less cellular. It expresses stronger and diffuse S100 positivity.

In most cases of SNF, complete surgical excision is the treatment of choice due to cosmetic concerns with extremely rare chances of recurrences.

So far, only one case has been reported as PNF in the post-auricular area with the right auricular deformity.[5]

We describe a distinct case of solitary neurofibroma, presenting with an atypical morphology with nodular clustering, in a young adult and a location not commonly associated with this type of tumour. Our case highlights the diversity of neurofibroma presentations and emphasises the need for comprehensive evaluation in unusual cases.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Okamoto M, Ueda T, Fukui M, Mitsui T, Masuoka H, Hihara M, *et al*. A Solitary Giant Neurofibroma of the Left Thigh: A Case Report. J Surg Case Rep 2022;2022: rjac359.
2. Sahoo A, Ali SM. Solitary Popliteal Neurofibroma: An Unusual Encounter. BMJ Case Rep 2020;13:e232771.
3. Kim KS, Lee DG, Lee DH, Hwang JH, Lee SY. Slowly Growing Solitary Neurofibroma of the Thumb: A Case Report. Medicine (Baltimore) 2021;100:e23611.
4. Choi JH, Ro JY. Cutaneous Spindle Cell Neoplasms: Pattern-Based Diagnostic Approach. Arch Pathol Lab Med 2018;142:958-72.
5. Wright EM, Sach E, Sharif S, Quarrell O, Carroll T, Whitehouse RW, *et al*. Can the Diagnosis of NF1 be Excluded Clinically? A Lack of Pigmentary Findings in Families with Spinal Neurofibromatosis Demonstrates a Limitation of Clinical Diagnosis. J Med Genet 2013;50:606-13.

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