Indian Journal of Pathology and Oncology 2023;10(3):313–316

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Indian Journal of Pathology and Oncology

Journal homepage: www.ijpo.co.in

Case Report

Neurofibroma of breast- A rare case report

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ARTICLE INFO

Article history:
Received 10-05-2023
Accepted 06-07-2023
Available online 27-09-2023

Keywords:
Neurofibroma
Nerve sheath

ABSTRACT

Neurofibroma are benign tumour of peripheral nerve sheath that are mostly sporadic. Solitary neurofibromas of breast are uncommon. In this study, a case of 21-year-old female was referred with complaint of hyper pigmented lump in left breast since birth with recent increase in size for 5 months. Ultrasonography revealed a well-defined hypoechoic Space occupying lesion in subcutaneous plane at 11'o clock position of left breast along with a large ill-defined hypoechoic area with normal vascularity noted extending from 11'o clock to 2'o clock areas in left breast. Microscopically, the underlying stroma showed proliferating spindle cells with interspersed collagen bundles. Spindle cells show bland serpentine (wavy dark) nuclei and scant cytoplasm.

1. Introduction

Neurofibromas are rare and benign tumours of peripheral nerve sheath which can be a common manifestation of neurofibromatosis type 1(NF1).[1] They usually consist of Schwann cells, collagen fibers and fibroblasts. They mostly occur on the trunk and limbs, usually occurring in almost 95% of individuals with NF1.[1] Neurofibromas are commonly found in the chest, back, elbows and knees in descending order. Nipple areola complex is the most common location of the breast neurofibroma. Mutation of NF1 gene leads to a non-functional neurofibromin protein that usually acts as a tumour suppressor. There are very few cases of breast neurofibromas that are reported. Almost 66 cases have been reported so far. It is an autosomal dominant condition which can present as multiple neurofibromas within the subcutaneous tissue.[1]

2. Case Report

A 22-year-old female came to Era Lucknow Medical College & Hospital. The female complained of hyperpigmented lump in left breast since birth with increase in size and pain along with increased itching and loss of appetite for 5 months. On physical examination, a single irregular shaped, mobile, hyperpigmented mass approximately 3.5 x 4 cm in size with no evidence of discharge, ulcer and fungation was seen in the upper inner quadrant, it was tender with normal temperature without any attachment to the underlying tissue. There was no evidence of any dilated engorged veins. Right sided central axillary lymph nodes were enlarged around 0.5 cm×0.5 cm. Few subcentimetric lymph nodes were noted on the left side.

The remaining physical examinations had no clinical findings. The neurological parameters were seen to be within normal levels. At the time of admission; the vitals of the patient were within normal range.

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https://doi.org/10.18231/j.ijpo.2023.070

Imaging involves MRI finding as an area of altered signal intensity with marked thickened skin and subcutaneous tissue medially in upper inner quadrant of left breast with evidence of a benign skin lesion. Ultrasonography findings suggest a well-defined hypoechoic space occupying lesion in subcutaneous plane at 11'o clock position of left breast with a large ill-defined hypoechoic lesion with minimal vascularity noted from 11'o clock to 2'o clock position in left breast. Mammography shows a homogeneous circumscribed mass with a regular border, without any calcification. Surgical excision was done for the condition under general anaesthesia. (Figure 1)

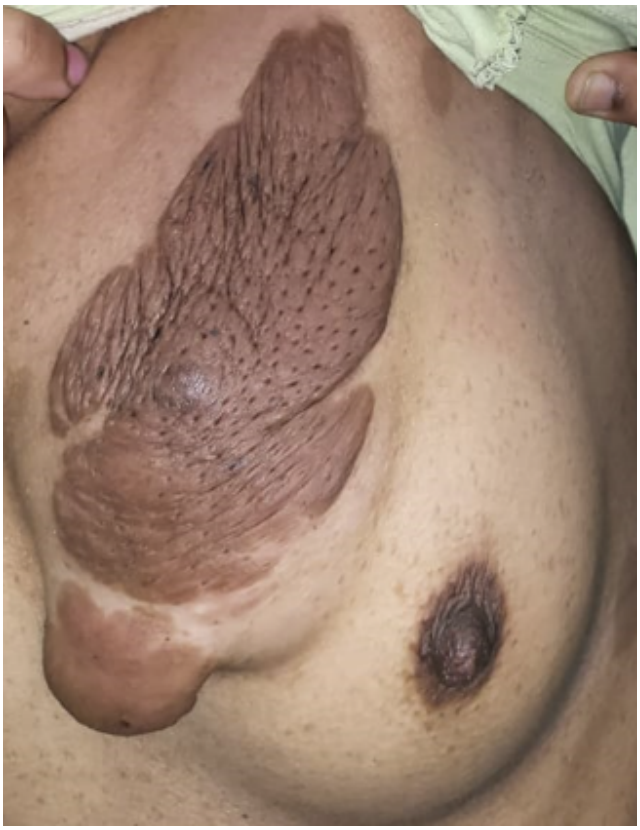

Fig. 1:

Gross- We received a skin covered specimen Gray-brown to Gray white, soft tissue piece measuring 13 x 7.5x 2 cm. Under surface shows fibrofatty tissue.(Figure 2)

Microscopy- Section from the tissue shows stratified squamous epithelium lining. The underlying stroma shows proliferating spindle cells with interspersed collagen bundles. These spindle cells have bland serpentine nuclei and scant cytoplasm. At areas, proliferation of small calibre blood vessels and necrosis seen. (Figures 3, 4 and 5)

3. Discussion

Neurofibromas are the benign slowly growing peripheral nerve sheath tumours. The most common presentation is

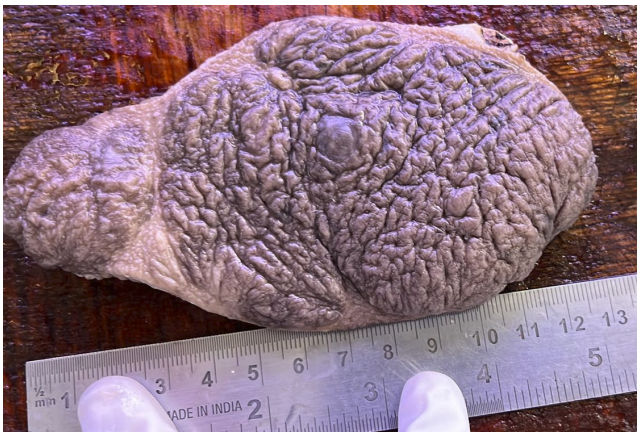

Fig. 2: Cut surface shows Gray brown areas

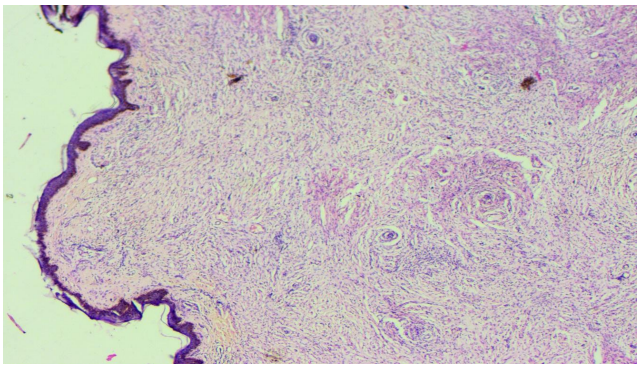

Fig. 3: Scanner(4x) Hematoxylin and Eosin- Stained Section shows proliferation of spindle cells Lined by stratified squamous epithelium

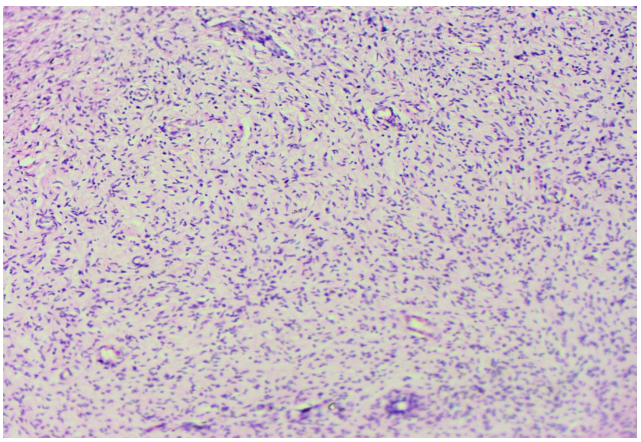

Fig. 4: Hematoxylin & Eosin-stained section in low power examination (10x) shows proliferation of spindle cells having bland serpentine nuclei and scant cytoplasm separated by collagen bundles.

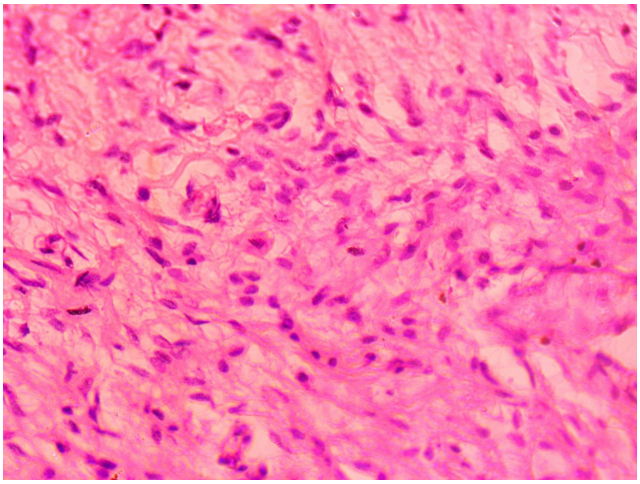

Fig. 5: Hematoxylin & Eosin-stained section in high power examination (40x) shows proliferation of spindle cells having bland serpentine nuclei and scant cytoplasm separated by collagen bundles

between age group of 20 and 30 years. However, no sex predilection can be seen.[2]

Presence of a tissue solitary mass within the dermis or in the subcutis is the commonest clinical presentation.[2] Localised, Diffuse and Plexiform are the three different varieties of the neurofibromas that have been described. Localised variety form approximately 90% of these lesions, and the vast majority are solitary and usually not associated with NF. Breast involvement is not very common, but the nipple-areola complex is the most common location in the breast. However, in this case, the tumour was seen in the inner side of upper quadrant of the left breast and not in nipple areola complex which is the commonest location of the neurofibroma of chest. On physical examination, cutaneous neurofibromas, appear as flesh coloured nodules or papules of different sizes. The size of the tumours may vary from several millimetres to a meter. They are either gelatinous in consistency or firm and nodular.[3] These may be pedunculated, dome shaped, or sessile and growth can cause cosmetic disfigurement.[3] As differentiated from plexiform neurofibromas which are usually present at birth, cutaneous neurofibromas generally develop after adolescence.[4] They usually course along the path of somatic, cranial, or autonomic nerves.[4] Most of these tumours are asymptomatic and are neither painful nor tender. The size of the tumours may vary from several millimetres to a meter. They are either gelatinous in consistency or firm and nodular.[4] They are usually aggravated by endocrine changes and are more commonly by trauma, menarche, or infectious diseases.[4] The pigmentary changes occur at birth, but the tumours do not appear until later in the first to second decade. They are usually called button defects because of their protrusion through a discrete dermal defect.[5] Various reports describe the sarcomatous change of neurofibromatosis incidence as being anywhere from 2 to 16%. Their malignant potential is usually associated with deep dermal tumours as opposed to those that are papillary or superficial.[5] The vast majority of tumors are located in the head, neck, and the chest wall areas.

Neurofibromas have been described as causing notable changes in bones, usually those that are caused by erosion.[6] Intraosseous cystic changes are also often noted. Other congenital problems such as bowing and scoliosis are common.[6] Endocrine changes have also been described. Acromegaly, cretinism and menstrual abnormalities are common. Infertility and Addison's disease are also correlated with the disease.[6] A curious association exists between neurofibromatosis and pheochromocytoma. Of patients with pheochromocytoma, 5 to 20% also have neurofibromas.

Microscopically, neurofibromas typically consist of spindle shaped cells having elongated wavy nuclei with no features of atypia or areas of high mitotic activity. Surgical excision is needed for the treatment of solitary neurofibromas.[6] Deep seated lesions and lesions involving major nerve trunks are preferably treated conservatively, given that surgery requires sacrifice of patient's nerve.[7]

Solitary neurofibromas usually have excellent prognosis. When not associated with NF-1, malignant degeneration is rare.[7] Local recurrence after excision is not common for lesions not associated with NF-1.[7] An important panel of Immunohistochemistry for the diagnosis of neurofibromas includes broad spectrum and basal cytokeratins.[7] IHC should be interpreted in the light of morphological findings as no single marker is fully specific.

4. Conclusion

This case with unilateral nipple areola complex neurofibroma presenting in a female displays an unusual presentation and diagnosis within histopathology with a vast array of differential diagnosis.[8] Immunohistochemistry plays a crucial role in the diagnosis of breast spindle cell lesions.[8] Solitary neurofibroma of breast is an uncommon benign tumour which can mimic other more aggressive tumours. Imaging plays an important role, however, diagnosis relies on pathology.[8] MRI is the best imaging technique to diagnose morphology and enhancement pattern suggestive of a benign lesion.[9]

5. Source of Funding

None.

6. Conflict of Interest

None.

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Cite this article: Shukla G, Bassi N, Irfan S, Zaidi N, Ahmad S, Lal N. Neurofibroma of breast- A rare case report. *Indian J Pathol Oncol* 2023;10(3):313-316.