Original Article

Prospective Study of Peripheral Nerve Tumors over a period of 2 Years

Suryakala Chappa, Sudhasmita Rout, K Prasad Reddy, PVB Ramalakshmi

Abstract : Peripheral nerve tumors arise from Schwann cells and perineural cells. Most of them are benign but have overlapping entities with different course of prognosis, recurrence and malignant transformation. The practicing pathologist must be familiar with their morphological features. We report 22 cases of Neural tumors, of which 11 were Neurofibromas, 10 were Schwannomas and 1 was Malignant Peripheral Nerve Sheath Tumor (MPNST).

Key Words : Neurofibroma, Peripheral nerve tumors, Schwannoma

Introduction

Peripheral nerve tumors arise from Schwann cells, Perineurial cells and Fibroblasts. Majority of them are benign but they have overlapping entities, different course of prognosis, recurrences and malignant transformation. The practicing pathologist must be familiar with their morphological features. An inherent challenge to the FNAC evaluation is that, there is cyto-morphological overlap between benign and malignant lesions. With FNAC, there is dispersion of individual cells and partial loss of recognizable tissue pattern, leading to less specific diagnosis. Our aim is to prove the efficacy of FNAC in the diagnosis of peripheral nerve tumors and its histological correlation.

Aims and Objectives

To identify the age, sex and site wise distribution of neural tumors, identify type of neural tumor and to correlate cytology with histopathology.

Materials and Methods

The present study is a prospective study of neural tumors conducted at the department of pathology, Maharajah's Institute of Medical Sciences, from Jan 2012 to Dec 2013. All patients of peripheral nerve tumors were subjected to FNAC followed by biopsy. Aspiration was done with 22G needle and 5cc syringe and smears stained with Hematoxylin-Eosin(H&E). For histopathology, the tissue was formalin fixed and paraffin embedded sections were cut and stained with H&E. Special stains and immuno-histochemistry (IHC) were done wherever necessary.

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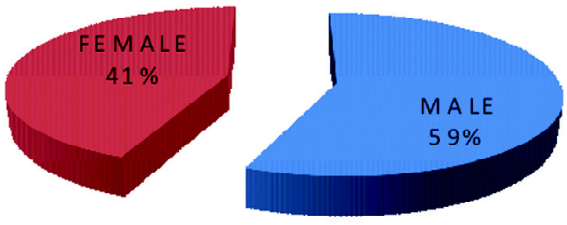
Results

Out of 167 cases of soft tissue tumors, 22 were peripheral nerve tumors. Among these 22 cases of peripheral nerve tumors 21 were benign and 1 was malignant. On FNAC all cases were reported as benign and on histopathology, 1 case was malignant.

Age Distribution

Parameter	Age (years)
Mean Age	38
Youngest age	11
Oldest age	76

Sex Distribution



Site Distribution

Site distribution	Number of cases
Head & neck	08
Lower extremity	07
Upper extremity	06
Trunk	01
Total	22

Types of Peripheral nerve tumors number

Neurofibromas	11
Schwannomas	10
MPNST	01
Total	22

Cyto-Histological Correlation

Cytology: Benign: 22, Malignant: 0.

Histopathology: Benign: 21, Malignant:1

Correlation = 95.45%

Discussion

Peripheral nerve tumors are classified into:

1) Benign: Schwannoma, Neurofibroma, Perineurioma
2) Malignant: MPNST (Malignant Peripheral Nerve Sheath Tumors)
3) Non-neoplastic: Traumatic neuroma and Morton's neuroma[1]

Schwannoma (Neurilemmoma) : Encapsulated tumor with yellowish cut surface with cystic degeneration(fig1). Mostly solitary but multiple and bilateral tumors can occur in association with NF2. Most common sites are head & neck, flexor aspects of upper and lower limbs. Other sites are mediastinum, retroperitoneum and cerebello-pontine angle.

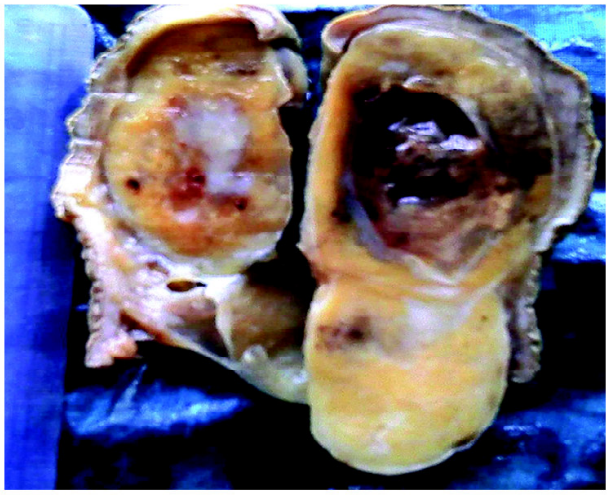


Fig 1. Schwannoma - gross. Note the well encapsulated tumor with yellowish discoloration and cystic changes

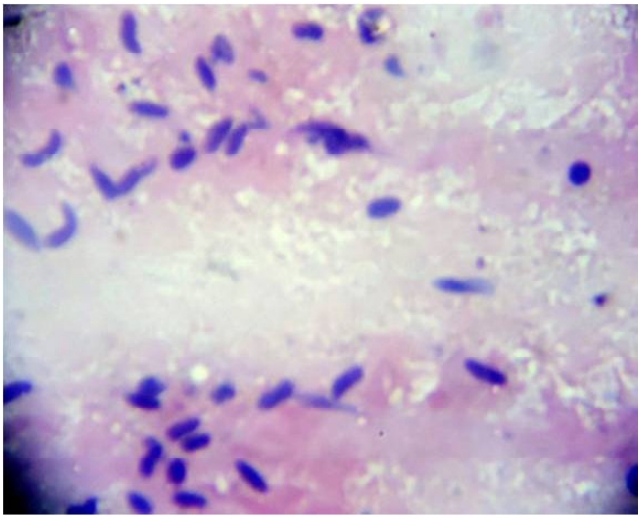


Fig 2. FNAC showing hypo-cellular smears with benign spindle cells

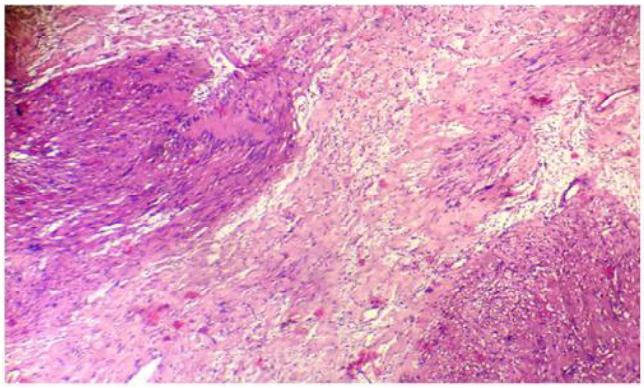


Fig 3. Schwannoma histopathology showing Antoni A and Antoni B areas

FNAC shows benign spindle cells and the smears are hypo-cellular (fig.2). Microscopy hallmark is the pattern of alternating Antoni A and B areas (fig3). Malignant transformation in Schwannoma is a very rare event unlike in neurofibromas. A rare variant of psammomatous melanotic Schwannoma is regarded as low grade malignancy because of its tendency for local recurrence.

Neurofibroma : Classified into

1) Localised: not associated with genetic syndromes
2) Diffuse: most common in head & neck as raised plaque lesion.
3) Plexiform: entire length of nerve is involved to appear as a bag of worms.

Diffuse and plexiform neurofibromas are associated with NF1.

Grossly appears as well-circumscribed, with cut surface yellowish and mucoid (fig4). Microscopy shows interlacing bundles of elongated cells with wavy dark nuclei (Fig5).

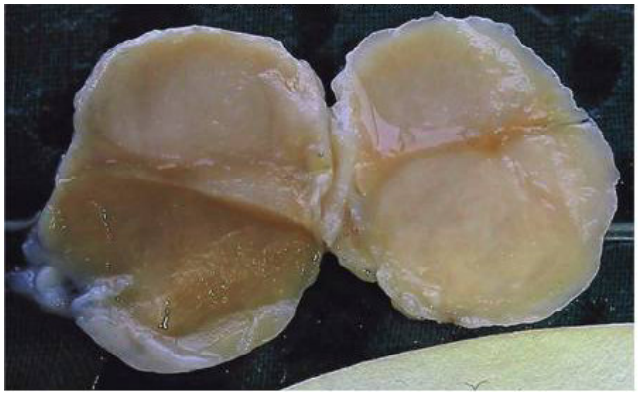


Fig 4. Neurofibroma – Gross well circumscribed yellowish white mucoid cut surface

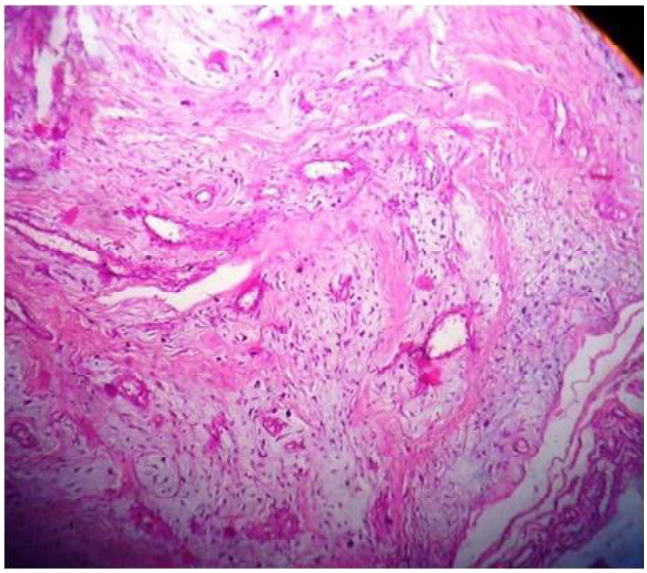


Fig 5. Histopatholgy neurofibroma showing Schwann cells, Fibroblasts and Nerve bundles

Neurofibromatosis (Von Recklinghausen disease)

Autosomal dominant. Type1 : NF1 gene is located on chromosome 17. It presents as plexiform neurofibromas (elephantiasis neuromatosa) with café-u-lait spots and Lisch nodules on the iris. Other associated features include megacolon, schwannomas, pheochromocytoma, neuroblastoma and GIST. Type2 : associated gene is NF2 is located on chromosome 22. It presents in CNS tumors like bilateral acoustic schwannomas, meningiomas, Optic nerve gliomas and astrocytomas. MPNST : Arise de-novo or part of Type 1 von Recklinghausens disease.

Neurofibromatosis should be suspected if tumor develops in Type 1 NF or tumor arising within the anatomic site of a major nerve. Grossly it is a large fusiform mass, fleshy, opaque, white-tan surface marked by areas of secondary hemorrhage and necrosis (fig6). Microscopy shows densely cellular fascicles alternate with hypocellular, myxoid zones, creating a marble-like effect (fig7). IHC of peripheral nerve tumors shows S-100 positivity (fig8).

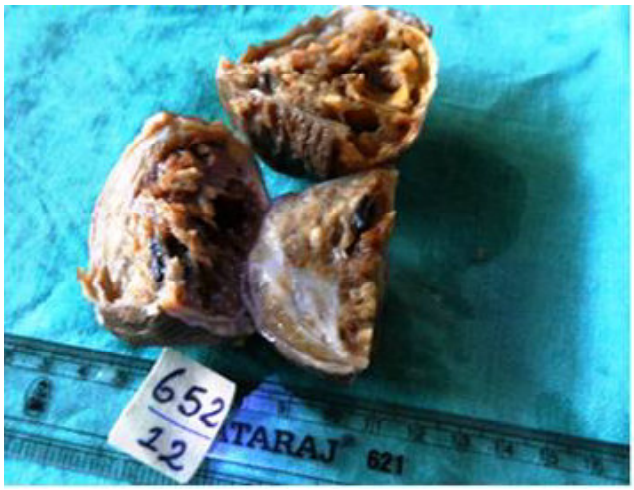


Fig 6. MPNST Gross section showing yellowish white masses with haemorrhage and necrosis

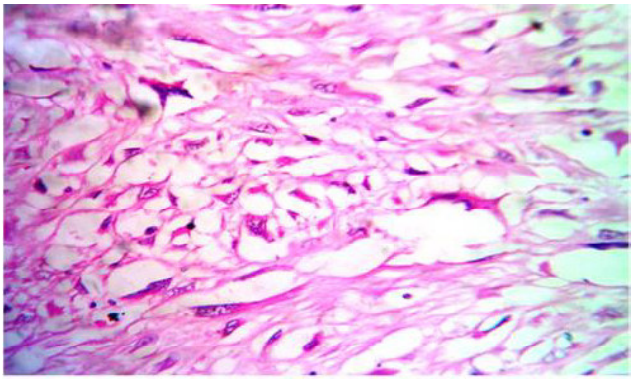


Fig 7. MPNST microscopy showing cellular areas with bizzare hyperchromatic nuclei

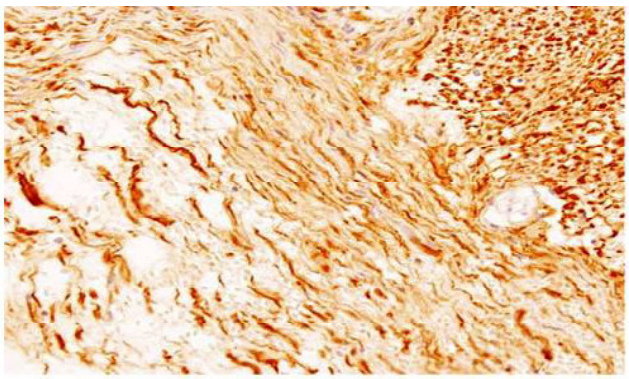


Fig 8. IHC for Schwannoma, S100 strong positivity

Conclusion

Most peripheral nerve sheath tumors in this study were benign. The mean age of patients was 36 years. The most common site was head and neck. Fine needle aspiration provided a relatively non-traumatic rapid diagnosis. Benign tumors ware easily diagnosed by FNAC, but benign cellular tumors and low grade malignancies needed confirmation by histopathology. The present study, whose aim was to prove the efficacy of FNAC as a useful tool and a reliable technique in diagnosing peripheral nerve tumors showed a sensitivity of 100% and specificity of 95.5%.

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