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Case Series

Role of S-100 immunocytochemistry in diagnosis of pure neuritic leprosy on fine needle aspiration cytology – A series of two interesting cases presented as nerve abscess

Subramanian Kalaivani Selvi[1], Khowsalya Subrajaa[2]*, Neelaiah Siddaraju[3]

[1]*Dept. of Pathology, Sri Venkateshwara Medical College, Hospital and Research Centre, Puducherry, India*
[2]*Dept. of Pathology, Bhaarath Medical College and Hospital (BIHER), Chennai, Tamil Nadu, India*
[3]*Dept. of Pathology, Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry, India*

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ABSTRACT

Pure neuritic leprosy (PNL) constitutes to 4.2 % of all leprosy cases in India. Histopathology is crucial for the diagnosis of PNL, but fine needle aspiration cytology (FNAC) is now replacing it, as nerve biopsy is a cumbersome procedure and can lead to neurological deficits. FNAC, a minimally invasive procedure can play a significant role in primary diagnosis of PNL. Apart from the cytomorphological features, using ancillary tests can further improve the diagnostic accuracy of PNL. We present here two cases, who presented with a soft tissue swelling. FNAC attempted from the lesions yielded a necrotic material which on microscopy showed abundant caseous necrosis with epithelioid cell granulomas and there were also few spindle shaped cells of neural morphology seen in clusters as well as scattered in the necrotic background. S-100 immunostaing was attempted which highlighted the neuronal cells confirming the diagnosis of PNL.

1. Introduction

Pure neuritic leprosy is a type of leprosy that involves only peripheral nerves and constitutes to 4.2% of all leprosy cases in India.[1] The common presenting symptoms are paresthesia, sensory or motor deficit and pain.[1] PNL presenting as a soft tissue swelling/ abscess is rare.[2] Diagnosis of PNL depends on the classical involvement of nerves that is demonstrated by clinical, electroneuromyographical findings and by morphological evaluation. Histopathological evaluation necessitates nerve biopsy which have limitations like difficult procedure and permanent nerve deficit. Fine needle aspiration cytology is now emerging as a nerve sparing diagnostic modality in PNL cases with less side effects. In addition performance of adjunctive tests on FNA material can increase the diagnostic accuracy. We present here two cases of PNL where in addition to cytomorphology, S-100 immunocytochemistry was performed to highlight the neuronal cells, thereby confirming the diagnosis of PNL.

2. Case Reports

2.1. Case 1

A 22 year old male patient presented to the surgery department with swelling over the left arm for two months which was diagnosed clinically as soft tissue swelling and was advised for FNAC. There was no history of any hypoaesthesia or any skin lesions. On examination there was a swelling in the subcutaneous plane measuring 3x1.5 cm over the posteromedial side of the left arm. The swelling was firm, nontender and was mobile only on horizontal direction. The vertical mobility was restricted suggestive of

* *Corresponding author.*
E-mail address: klvnselvi@yahoo.co.in (K. Subrajaa).

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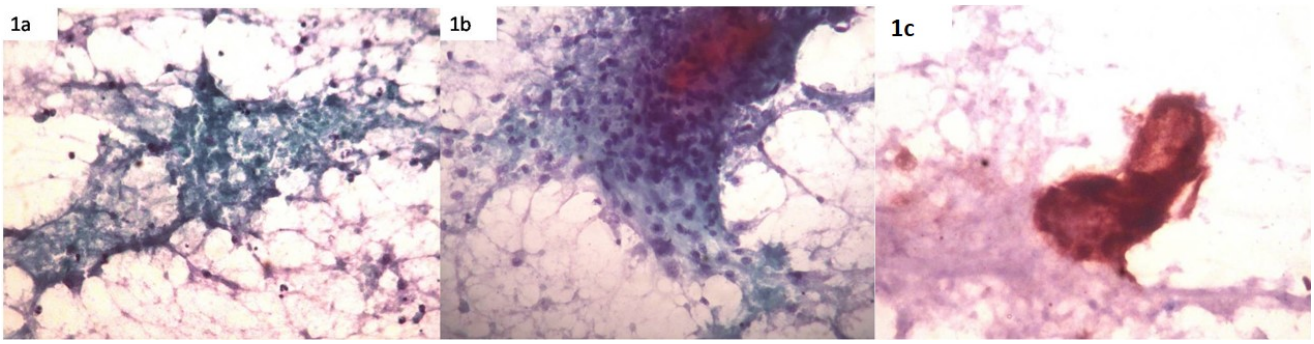

Figure 1: a: Cytological examination showingcaseous necrotic material (PAP, 400X); **b:** Epithelioid cell granuloma (PAP, 400X); **c:** Immunocytochemistry with S100 highlighting the neural cells in the nerve twig (Immunocytochemistry, 400X).

neural origin. FNAC was attempted from the swelling which yielded thick pus like necrotic material. Microscopy showed abundant caseous necrosis (Figure 1a) with epitheloid cell granulomas (Figure 1b) and also there were few spindle cells arranged in clusters as well as scattered in the background resembling neural origin. Stain for acid fast bacilli was negative. Correlating with the clinical features and the FNAC findings a diagnosis of tuberculoid neuritic leprosy was considered. However we attempted to highlight the neuronal cells using S-100 immunocytochemistry which had stained the spindle cells confirming the neural origin (Figure 1c). The patient was then referred to dermatology for further treatment.

2.2. Case 2

A 31 year old male patient present with swelling in the foot . No other dermatological lesions detected. The swelling was firm and mobile in the horizontal plane with restricted vertical mobility. FNAC from the swelling yielded pus like material. Cytological features showed caseous necrosis with epithelioid cell granulomas. Acid fast stain was negative. S-100 stain again highlighted the neuronal cells. Thus a diagnosis of tuberculoid PNL was made and refered the patient to dermatology department.

3. Discussion

Pure neuritic leprosy ia an inflammatory disease of nerves without cutaneous involvement, and presents commonly as peripheral neuropathy. Diagnosis of PNL is usually based on the 1) Demonstration of thickened nerves, altered sensation, atrophy or hypertrophy of muscles causing functional disability; 2) Demonstration of perineural inflammation or neuritis in nerve or skin biopsies. 3) Demonstration of acid-fast bacilli in nerves. 4) PCR confirmation for DNA of M. leprae. 5) Detection of anti-GLP-1 IgM antibodies. 6) Altered electromyography (EMG) when other evidences are absent to explain the alteration.[3]

As PNL lacks classic dermamatological lesions, the diagnosis mostly warrants histopathological examination of nerve biopsies to differentiate from other causes of peripheral neuropathy which has drawbacks like permanent nerve deficit, sampling difficulties and low sensitivity. Skin biopsies also fail to demonstrate histopathological changes of leprosy.[4] Fine needle aspiration cytology is now emerging as a nerve sparing technique in PNL with high diagnostic yield with the added advantage of less side effects.

Apart from diagnosis FNAC can also help in classification of leprous neuritis according to Ridley-Jopling scale from tuberculoid to lepromatous pole.[2,5,6]Tuberculoid PNL usually presents with epithelioid granulomas with or without caseous necrosis. Caseous necrosis in leprosy is frequently associated with tuberculoid PNL wheras the cutaneous tuberculoid forms of leprosy is never associated with caseaus necrosis.[5,6]

Mehdi G et al. was able to diagnose leprosy on cytology in 92 % of their cases. They were also able to classify them according to Ridley-Jopling scale as tuberculoid and lepromatous types.[7] In a study by Rao S et al., cytological diagnosis of lepsrosy correlated well in tuberculoid (90%) and in lepromatous (93.7%) cases than borderline cases when compared with histopathological evaluation.[8] Siddaraju et al have also highlighted the role of FNAC in diagnosis of PNL that presented as nerve abscess.[2]

When a single nerve is involved, the differential diagnosis includes nerve sheath tumors like neurofibromas and schwannomas, sarcoidosis, and sporotrichosis.[9] And with presence of granulomas sarcoidosis and sporotrichosis are the close differentials. Sarcoidosis can be differentiated by absence of necrosis and inflammatory cells as it usually shows only naked granulomas. Sporotrichosis can be identified by the presence of fungal elements and it ussualy shows suppurative granuloma with plasma cells. Presence of frank caseous necrosis with epithelioid granulomas should raise the suspicion of PNL.

Schwann cells can cytologically resemble epithelioid cell granuloma. Kumar et al have also described the presence of schwann cells in the aspirate from PNL lesions and discussed about the difficulties in differentiating it from the epithelioid cells.[9] Kaushik et al have also reported the presence of schwann cells in the aspirate.[10] S100 the marker of schwann cells can highlight those cells and can differentiate from the epithelioid cell granulomas. In these two cases S-100 immunocytochemistry was used to highlight the schwann cells which in correlation with cytomorphological features and clinical history strongly favoured the diagnosis of PNL.

S-100 is a commonly used immunomarker and is available in most of the laboratories and can be used to increase the diagnostic accuracy of PNL.

4. Conclusion

FNAC is a nerve sparing procedure that can replace biopsy in the diagnosis of PNL especially when presenting as nerve abscess and the use of S-100 immunostaining can increase the diagnostic accuracy of PNL confirming the neural origin.

5. Source of Funding

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6. Conflict of Interest

None.

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Author biography

Subramanian Kalaivani Selvi, Associate Professor

Khowsalya Subrajaa, Assistant Professor

Neelaiah Siddaraju, Professor

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