

Rare Neurogenic Tumor -Pacnian Neurofibroma: A Case Report

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ABSTRACT

Pacnian neurofibroma (PN), a rare benign dermal tumor characterized by proliferation of pacinian corpuscles in the dermal myxoid stroma. A 50 year female presented with slow growing, soft, painless swelling over right wrist. No other significant history was found. Histopathology revealed proliferation of fibrous connective tissue along with whorled structures. No cellular atypia was found. Immunohistochemistry examination with antibodies against protein S-100 showed diffuse positivity. A diagnosis of Pacinian neurofibroma was made.

Keywords: Pacinian Neurofibroma, Pacinian Corpuscles, S-100 Antibodies

Introduction

Pacnian neurofibroma (PN), a rare benign dermal tumor characterized by predominant proliferation of pacinian corpuscles in the dermal myxoid stroma. Few cases have been reported in the literature till now.^[1]

The term pacinian neurofibroma was initially described by Thoma in 1894, and later by Prichard and Custer in 1952.^[2] It usually presents as solitary nodule. Most common sites are fingers, hands, and feet, where the pressure receptors are usually located. However, these lesions are known to occur elsewhere also. Histopathological features usually show typical round to ovoid corpuscles, with multiple concentric lamellae in myxoid matrix.^[1,3]

We report a case of pacinian neurofibroma presented with painless wrist swelling.

Case Report

A 50 year old female presented with a slow growing painless right wrist swelling over dorsum aspect. There was no history of trauma. No other significant history was found. Physical examination revealed a soft, non tender swelling measuring 2.5x 1.5 cm. The patient underwent excision of the swelling.

Grossly, we received a skin covered, globular capsulated tissue piece measuring 2.2x1.5x1.2cm. Overlying skin cover measuring 2.2x1.5cm. On cut section homogenous yellowish white resembling adipose tissue.(Figure 1)

Microscopic examination revealed a well demarcated encapsulated dermal lesion composed of proliferation of variable sized concentric lobules resembling pacinian corpuscles. These concentric lobules merged with adjacent collagen fibres of dermis. The background show interlacing

bundles of spindle cells with wavy nuclei. At places, myxoid change seen in collagenous stroma. There was no evidence of cellular pleomorphism or atypical mitotic figures.(Figure 2,3) Immunohistochemistry was performed and showed strong diffuse positivity to protein S-100. (Figure 4) Based on morphological features and IHC, a final diagnosis of Pacinian Neurofibroma was made.

Discussion

Pacnian corpuscles are specialized nerve endings with a lamellated structure and are regarded as pressure and vibration receptors. They are largest sensory nerve-end organs located in the deep dermis and subcutaneous tissue and function as tactile receptors.^[4] Solitary tumors of peripheral nerve are relatively uncommon in the hand, representing less than 5% of all hand tumors. Benign tumors include schwannoma(neurilemmoma) and solitary neurofibroma. Pacinian neurofibroma is a rare variant composed of structures resembling pacinian corpuscles at various stages of maturation. Well differentiated Pacinian corpuscle shows homogenous, acellular, eosinophilic central core, while more immature stage contains more cellular elements with spindle shaped nuclei.^[5]

Thoma in 1894, first described the term Pacinian neurofibroma and later by Prichard and Custer in 1952 as well as by Prose *et al*, in 1957. Pacinian hypertrophy, pacinian hyperplasia and neurofibroma should be considered in differential diagnosis. Pacinian hyperplasia exhibits hamartomatous overgrowth and is usually associated with history of prior trauma. The classical structure of pacinian corpuscles is well maintained both in pacinian hypertrophy and hyperplasia.

Pacnian neurofibroma, on the other hand shows pacinian corpuscles-like differentiation in various stages of

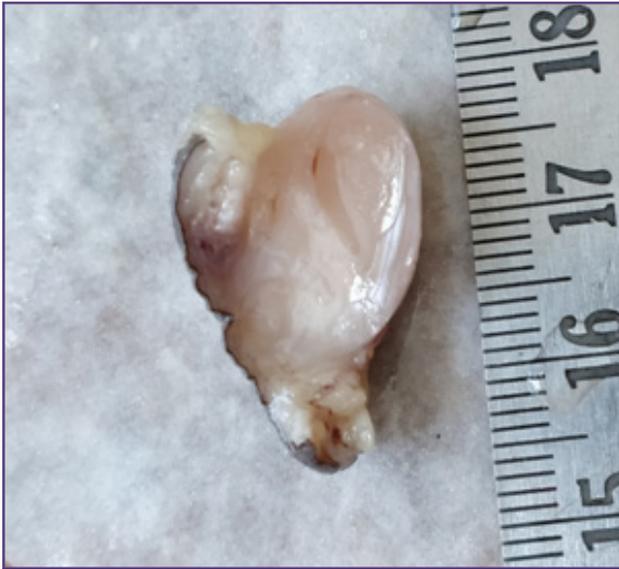


Fig. 1 Grossly, a globular skin covered tissue piece.

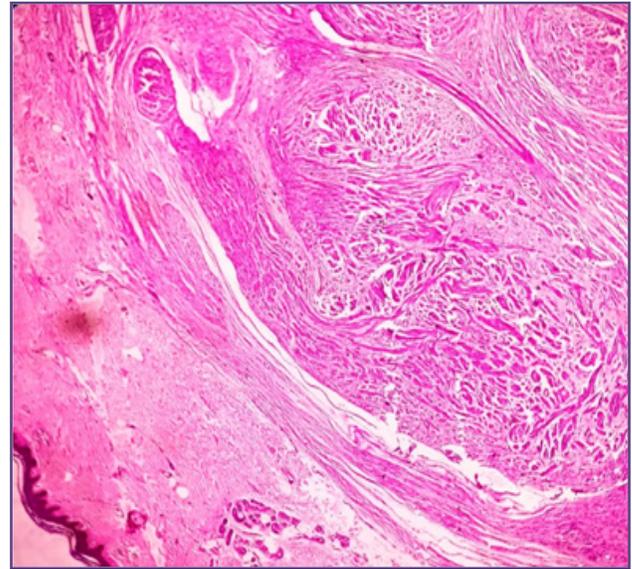


Fig. 2 Photomicrograph showing stratified squamous epithelium with subepithelium showing pacinian corpuscles. (H&E 20x).

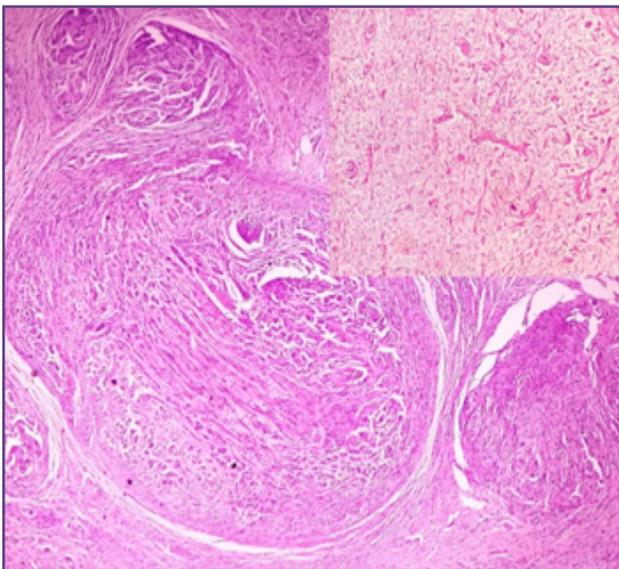


Fig. 3 Photomicrograph showing pacinian corpuscles with different stages of maturation (H&E 20x). Inlet box shows myxoid stroma at places.

maturation within a myxoid stroma. No association with neurofibromatosis has been found. Ordinary neurofibroma may show occasional mature pacinian corpuscles, but different stages of maturation are not present. The presence of multiple neurofibromas is associated with neurofibromatosis.^[6]

Pacinian neurofibroma is a rare benign dermal tumor, usually presents with solitary, soft to firm, well demarcated,

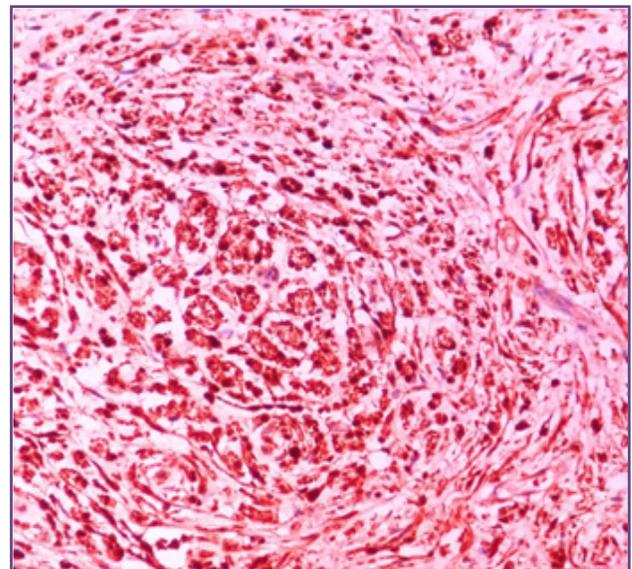


Fig. 4 Immunohistochemistry showing strong positivity against S-100.

mobile nodule as in our case. Multiple lesions are rare. Most commonly they occur on hands and feet but other reported sites are buttocks, neck, flank, arm, cheek and sacro-coccygeal region.^[7] They are usually seen in adolescents and young adults but our case presented in fifth decade.

There is variation in clinical presentation, appearance and histology of the tumor. Previous literature describe it as a painful, firm, solitary dermal nodule on middle-aged adult;

some report a history of trauma.^{[1] [8]} Our case presented with soft, painless swelling.

Histopathology has a critical role in confirming diagnosis because clinical features are less typical in most cases. In our case histopathological features include a well demarcated lesion with round to ovoid lobules that contain pacinian corpuscles embedded in fibrous connective tissue stroma. At places, depicting myxoid change. Immunohistochemistry showed tumor cells with positive immunoreactivity against antibodies S- 100.

Pacinian neurofibroma is not found to be associated with von Recklinghausen's disease or any other syndrome which requires additional workup or surgery. There is no evidence in the literature that these tumors have a high rate of recurrence or any tendency to undergo malignant change. The variation in clinical presentation may make preoperative diagnosis difficult and the final diagnosis may lie in the hands of the pathologist.^[9]

Wide surgical excision is the treatment of choice after assessing various factors e.g size of tumor mass involvement of vascular and nerve structures.

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