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NEUROTIZED CONGENITAL MELANOCYTIC NEVUS OF THE NONGIANT TYPE.

Pathology

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ABSTRACT

Introduction: A congenital melanocytic nevus (CMN) is a lesion present at birth and containing nevus cells. Congenital nevi are rare and found in about 1% to 2% of newborn infants and are usually solitary. The appearance varies considerably on the basis of morphology, texture, location, and size. Clinically, congenital melanocytic nevi are classified on the basis of their size. Large or giant congenital melanocytic nevi are those which are 20 cm or more in greatest diameter. The term neurotization refers to the resemblance of some melanocytic cells to peripheral nerve sheath cells. In neurotization there is presence of elongated or slender melanocytes (exaggeration of the type C melanocytes) with fibrillary cytoplasm, often in the mid to deep portions of some nevi. Although neurotisation may be seen in giant congenital melanocytic nevi, it has not been reported in the Nongiant type.

KEYWORDS

Congenital melanocytic nevus, Nongiant, Neurotized

INTRODUCTION:

A congenital melanocytic nevus (CMN) is a lesion present at birth and containing nevus cells. Congenital nevi are rare and found in about 1% to 2% of newborn infants.[1] Although neurotisation may be seen in giant congenital melanocytic nevi,[2,3,4,5] it has not been reported in the Nongiant type.

Case Report:

A 20 year female presented with pigmented lesions on the left thigh and flank region. They were present since early childhood, and progressively increasing in size. The lesions were not associated with pain or itching. There was no family history of a similar skin disorder. Patient wanted the lesions to be excised for cosmetic reasons.

On examination, the lesion in the flank region was ill-defined and spread over an area of 15 cms. There were areas of hyperpigmentation. The skin was intact and there was no tenderness. The thigh lesion was 3 cms in size. There were no other skin lesions. The lesion was excised and sent for histopathological examination. Grossly the excised skin showed a poorly defined macular hyperpigmented lesion measuring 15 cms in greatest dimension (Figure 1).

Figure 1 showing the excised specimen with an ill-defined hyperpigmented lesion.

On microscopic examination, the epidermis showed mild acanthosis. The dermal region showed diffusely scattered groups and nests of uniform round to oval cells having vesicular nuclei, and moderate amount of cytoplasm containing brownish-black pigment resembling melanin pigment. The cells in the upper portion showed abundant pigment, with decreasing amount of pigment in the deeper portions. Junctional activity was not seen. Heavily pigmented melanophages were seen in the surrounding tissue. The groups and nests of cells were seen in the deep dermis and the superficial subcutaneous tissue (Figure 2, 3). The underlying dermal tissue showed cells having wavy elongated nuclei resembling neural cells with fibrillary cytoplasm (Figure 4). These areas resembled a neurofibroma. A diagnosis of Neurotized Congenital Melanocytic Naevus was made. As the lesion was less than 20 cms, it was considered to be the nongiant type. The resected margin was free of cells and the excision was complete.

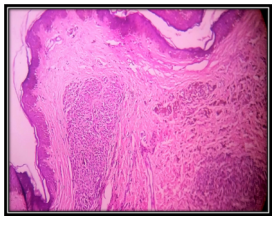

Figure 2: Showing clusters of nevus cells (H&E 10X).

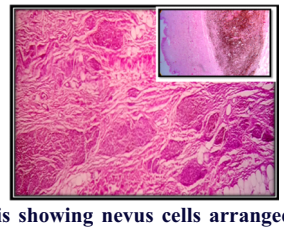

Figure 3: Dermis showing nevus cells arranged in nests. Inset; shows heavily pigmented melanophages (H&E 40X)

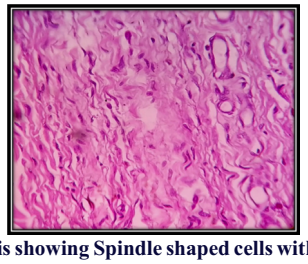

Figure 4: Dermis showing Spindle shaped cells with elongated and wavy nuclei (H&E 40X).

Discusssion:

Congenital nevi are usually larger than acquired nevi. However, only a few are of considerable size. Those measuring more than 20 cm in greatest diameter are referred to as *giant congenital melanocytic nevi.* The others are referred to as *nongiant congenital melanocytic nevi.* These are slightly raised, often pigmented lesions, and they may show a moderate growth of hair. They may be classified as "small" (less than 1.5 cm in diameter) or "intermediate" (greater than 1.5 cm and less than 20 cm).[1]

Nongiant congenital nevi differ from acquired nevi in terms of their greater size and depth and in the involvement of skin appendages and sometimes even deeper tissues. Nongiant congenital nevi may be junctional, compound, or intradermal nevi, and their location in the dermis may be either superficial, which may include junctional involvement, or both superficial and deep. Cytologically, the cells of congenital nevi are similar to those of acquired nevi.[1]

Giant Congenital Melanocytic Nevi are often are more complex than nongiant congenital nevi. When the neural nevus component predominates, formations such as neuroid tubes and nevic corpuscles are present. These areas may show considerable similarity to a neurofibroma.[1] The term neurotization refers to the resemblance of some melanocytic cells to peripheral nerve sheath cells. In neurotization there is presence of elongated or slender melanocytes (exaggeration of the type C melanocytes) with fibrillary cytoplasm, often in the mid to deep portions of some nevi. The melanocytes within neurotized areas are arranged in short bundles or structures resembling Meissner corpuscles or Verocay bodies. Some authors prefer to use the term melanocytic nevi with peripheral nerve sheath differentiation.[6,7]

Neurotized congenital melanocytic nevus and pigmented neurofibroma (PNF) are close mimics and pose a clinicopathological challenge. Melanocytic nevus and neurofibromas have a common origin, i.e. neural crest derived stem cells.[4,6] Neurofibromas are composed of a proliferation of all elements of a peripheral nerve, including Schwann cells, axons, perineurial cells, and fibroblasts. These are intermixed with a rich network of collagen fibres, numerous mast cells, and sometimes, mucinous change in the background. Neurotized melanocytic nevi are a subset of melanocytic nevi with areas composed of spindle-shaped melanocytes arranged in cords or fascicles resembling neuroid structures in the dermis. Neurotization involves only dermal melanocytes.[6,7]

Pigmented neurofibroma is a rare neurofibroma with melanin-laden pigment cells which accounts for less than 1% of neurofibromas. On histopathology, there is prominent clustering of nests or cords of melanocytes in CMN unlike PNF. Spindled cells with peripheral nerve sheath differentiation including Wagner-Meissner corpuscle like bodies and junctional and/or dermal melanocytosis are visible in both CMN and PNF. Prominent abnormal nerve structures and plexiform foci are seen in PNF not in CMN.[4,7] Our case showed naevus cells with a progressive maturation into spindle cells and differentiation into neuroid structures and hence a diagnosis of neurotized congenital melanocytic nevus was made.

The incidence of melanoma in nongiant congenital nevi is unknown, but it is thought to be greater than that in a comparable area of normal skin. It is likely that the risk is related to the lesion's size. The excision of all nongiant congenital nevi, where feasible, is advised by many although not all authors. Most of the melanomas observed in these nongiant lesions occurred after puberty.[1]

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