

Melanotic Neurofibroma: A Case Report

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Abstract

Introduction: Melanotic neurofibromas are rare tumours, derived from peripheral nerve sheath, whose originality consists in the presence of melanic pigment. The clinical and histological diagnosis is often difficult to make requiring the immunohistochemical exam to make the difference between melanotic neurofibroma and the other pigmented tumors. Melanotic neurofibroma has a good prognosis and the malignization is rare.

Case Report: A 22 year-old man presented since childhood a left occipital tumor, it had a firm consistency and pigmented color. CT objectified isodense occipital mass, which enhanced heterogeneously after contrast product. Lack of bone loss beside. Complete surgical excision was performed. Histological and immunohistochemical analysis led to the diagnosis of melanotic neurofibroma.

Discussion: Pigmented neurofibroma is a rare variant of neurofibroma showing melanin production and constitute less than 1% of all cases. It can occur on their own or be associated with neurofibromatosis. They must be distinguished from classical neurofibromas when pigmentation occurs in the latter. Melanotic neurofibromas usually appear in the second or third decade of life and rarely in childhood. The clinical diagnosis is difficult to establish requiring the histopathological examination to differentiate between the melanotic neurofibroma and other pigmented tumours. It can mimicking a giant naevus or a neurocristic cutaneous hamartoma. These are the two main differential diagnoses among children. Among adults, the main difficulty is to distinguish melanotic neurofibroma from pigmented dermatofibrosarcoma. The elective treatment is surgical, represented by the complete excision of the tumor.

Conclusion: Pigmented neurofibroma is a unique subtype of neurofibroma which is rare and contains melanin producing cells. The histopathological features with unique pattern of melanogenesis, ultrastructural findings and immunohistochemistry will enable us to diagnose this entity and differentiate it from other pigmented tumours of skin.

Keywords: Pigmented; Immunohistochemistry Melanotic; Neurofibroma; Skin; Histopathology;

Introduction

Pigmented or melanotic neurofibroma is a rare variant of neurofibroma. It is derived from peripheral nerve sheath, whose originality consists in the presence of melanic pigment. The clinical diagnosis is difficult to establish. So a histopathological exam is mandatory to make the diagnosis. Although, sometimes neither the anatomopathological exam can establish the final diagnosis, requiring immunohistochemical supplementary studies to rule out other differential diagnoses. Melanotic neurofibroma has a good prognosis. The elective treatment is surgical, represented by the complete excision of the tumor. We present a case of melanotic neurofibroma of the scalp in a young man of 22 years. We will study the clinical, histological, immunohistochemical and differential diagnosis of this rare benign tumor.

Case Report

A 22 year-old man without disease history. He presented since childhood a left occipital tumor, it increases gradually in volume. Clinical examination revealed a subcutaneous nodule mobile relative

to the two plans, it had a firm consistency and pigmented color. CT objectified isodense occipital mass, which enhanced heterogeneously after contrast product. Lack of bone loss beside. It was initially considered to be a congenital naevus.

Complete surgical excision was performed. Histological analysis showed a proliferation of spindle-cells within the dermis and subcutaneous layers, with multiple foci of melanin-laden cells but no mitotic figures or atypical cells. There was no melanocytic theca. The tumour had immunoreactivity for the S-100 protein and HMB-45. The association of a benign pigmented tumour producing melanin and the presence of nervous cells, led to the diagnosis of melanotic neurofibroma (Figure 1a-1d).

Discussion

Melanotic neurofibroma also named pigmented neurofibroma is a rare tumor, representing less than 1% of all neurofibromas. It can occur in patients with neurofibromatosis or be isolated. In a study of 17 patients who had one or more melanotic neurofibromas, 8 of them had neurofibromatosis type 1 [1]. It is a benign spindle cell tumor containing melanin granules. This tumor is more frequent in black race populations with an age range of 2-71 years but it is usually appear in the second or third decade of life and rarely in childhood, as our

patient was in the twenties years [2]. Melanotic neurofibroma, like the classic neurofibroma, usually involve head and neck, as our case, trunk, buttocks and legs, very rarely on the palms and plants [1].

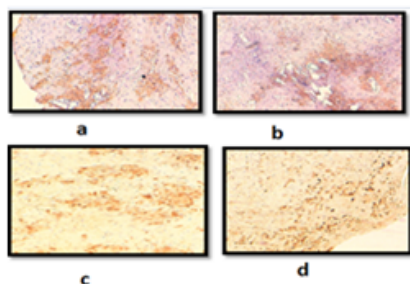


Figure 1: a) and b) spindle cells proliferation, with multiple foci of melanin-laden cells, C) immunostaining of HMB45+, d) immunostaining of PS100+.

Macroscopically, its size varies widely, from less than one centimeter in diameter to fifty centimeters. Its appearance is usually nodular, it is of elastic consistency and sometimes tends to invaginate to pressure [3-5]. It usually present a brown coloration more or less dark, sometimes blue, usually heterogeneous in relation to the pigment located deep in the dermis, whereas the classical neurofibroma is flesh-colored to light brown, secondary to increasing the pigment content in the basal level [4]. On our case the patient presented a subcutaneous nodule mobile relative to the two plans, it had a firm consistency and pigmented color.

Histologically, melanotic neurofibroma, like all neurofibroma, tumors of peripheral nerve sheath corresponding to a proliferation of fibroblasts and Schwann cells, associated with the nerve fibers, and located in the dermis and hypodermis [1,5]. Its originality is the presence of abundant melanin pigment within the neurofibromatous proliferation. It is contained in the form of granules intracytoplasmic in dendritic cells, spindle or more rounded cells, which tend to cluster, especially in the deep dermis. According to Motoi and al this unique distribution pattern of melanin producing cells in pigmented neurofibromas can be a major useful tool for distinguishing this tumour from other pigmented tumours [4]. We don't found atypia, mitosis or necrosis.

Immunohistochemically, the spindle cells are positive for S-100 protein and intratumoral melanogenesis is demonstrated by positivity to Melan-A, and HMB-45, markers specific for melanin granules [1,3].

Pigmented neurofibroma must be distinguished from other pigmented tumors. Among children, the main differential diagnosis is congenital nevi, so the melanotic neurofibroma differs from congenital nevi [6-8] by the absence of superficial pigmentation and thecae. Another differential diagnosis is pigmented dermatofibrosarcoma or Bednar tumor, which is an uncommon tumour with a predilection for the truncal region which is not usual for pigmented neurofibroma. It is important to distinguish the pigmented dermatofibrosarcoma of melanotic neurofibroma because the prognosis and the treatment are different [3,7]. The pigmented dermatofibrosarcoma is more densely cellular, often organized according to a storiform pattern and strongly positive for CD 34 and lacking of S-100 protein immunoreactivity. [3,9] Cellular blue nevus presents typically in young adults in the lumbosacral and buttock region. Sometimes one can observe this

lesion in children. The melanin pigment located deep in the dermis also gives it a bluish coloration. Histologically, cellular blue nevus consists of pigmented dendritic cells and fusiform cells organized in nodular pattern. This proliferation is denser and nodular than melanotic neurofibroma [8].

Melanotic schwannoma can be differentiated from pigmented neurofibroma by the presence of cellular areas alternating with hypocellular areas and dense area. The cells are slightly larger and epithelioid than the melanotic neurofibroma. They have a large nucleus with prominent nucleoli and are increasingly available palisade. The vessels have a thick wall. Moreover, one can see calcification and bone metaplasia in schwannoma, not observed in pigmented neurofibroma. Marking by the PS 100 is diffuse and often more intense. But the neurofilament and Synaptophysine are negative [1,5].

The elective treatment is surgical, represented by the complete excision of the tumor. Because of the diffuse pattern of growth these lesions may recur, but malignant transformation or metastasis has not been known [1].

Conclusion

Pigmented neurofibroma is a unique subtype of neurofibroma which is rare and contains melanin producing cells. The histopathological features with unique pattern of melanogenesis, ultrastructural findings and immunohistochemistry will enable us to diagnose this entity and differentiate it from other pigmented tumours of skin.

Consent from the Patient

"Written informed consent was obtained from the patient's legal guardian(s) for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal."

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