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Case Report

Treatment and Histopathology of Giant Congenital Nevocytic Nevi: An Exceptional Case of Neurotized Melanocytic Nevi

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Abstract

A 7-year-old female, without a medical history, she was noted at birth to have a confluent area of pigmentation over the proximal arm extending on to the axilla. The neoplasms had existed for 7 years without change. There was no history of pain, itching or discomfort. The concern of the present problem of the patient was only esthetic. Clinically, a pigmented nevus or neurofibromas was suspected. We have now undergone excision and reconstruction of the upper arm with skin grafts. An excisional biopsy was performed after operation. The diagnose of histopathology is neurotized melanocytic nevi. This case is reported because of its rarity.

Keywords: Giant Congenital Nevocytic Nevi; Histopathology; Immunohistochemistry; Neurotized Melanocytic Nevi;

Introduction

Giant Congenital Nevocytic Nevi (GCNN) are common benign cutaneous neoplasms, with a diameter greater than 20 cm, or may be greater than 2% of the body surface area. GCNN are congenital and could involve anywhere of the body surface area even the placenta. They are often round or oval, clearly demarcated and some times slightly intact. Pigmentation is usually even, although some congenital nevi have a speckled appearance. The course of GCNN may be varied. It may increase in size during childhood, show pigmentary regression with increasing age, develop a halo of hypopigmentation, or may spontaneously regress after halo formation or develop into malignant melanoma.

The cause of GCNN is not known, may be many factors□heredity [1]. GCNN are less common, relatively rare lesions with an incidence of 1 in 20000 live births. There is an associated increased risk of the development of malignant melanoma to be 8.5% during the first 15 years of life [2]. Three different patterns of GCNN may be seen: compound or intradermal nevus, 'neural nevus', and blue nevus pattern [3]. 'neural nevus'are rare.Toussaint S reviewed a large series of different types of melanocytic nevi and found neurotized melanocytic nevi was seen in 2 cases(0.1%) in all 2146 compound melanocytic nevi [4].

Case Report

A 7-year-old female, without a medical history, she was noted at birth to have a confluent area of pigmentation over the proximal arm extending on to the axilla (Figure 1). Opposite site right upper lime of the patient is normal skin. Wigth of patient is 37kg.Round size of the both upper lime is same 19cm.

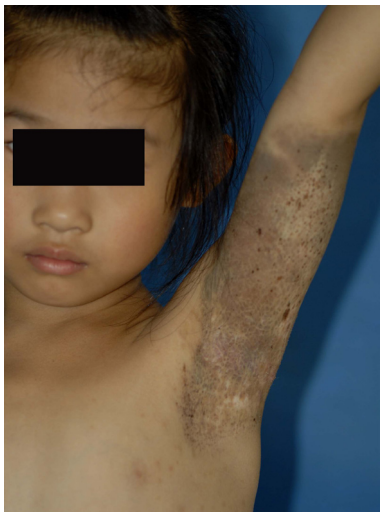

Figure 1: preoperative.

The neoplasms had existed for 7 years without change. No other family members were similarly affected. There was no history of pain, itching or discomfort. The concern of the present problem of the patient was only esthetic. Clinically, a pigmented nevus or neurofibromas was suspected. We have now undergone excision and reconstruction of the upper arm with skin grafts. Tissue expansion is now recognized as an invaluable tool in the treatment of giant nevi, but its application is limited in this case when there is limited normal tissue to expand, because the neoplasms encompass almost entire circumference of the arm.

An excisional biopsy was performed after operation. The diagnose of histopathology is neurotized melanocytic nevi (Figure 2).

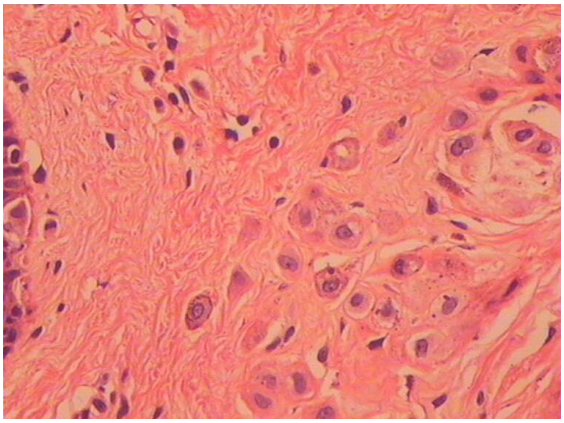

Figure 2: Histopathology of neurotized melanocytic nevus with nests of spindled melanocytes resembling Meissner corpuscles, in the mid dermis showing neurotization. (hematoxylin-eosin, original magnifications ×20).

The outcomes of immunohistochemistry is S-100 (+), Melan-A (+), HNB-45 (-), NF (-), Syn (-) (Figure 3,4).

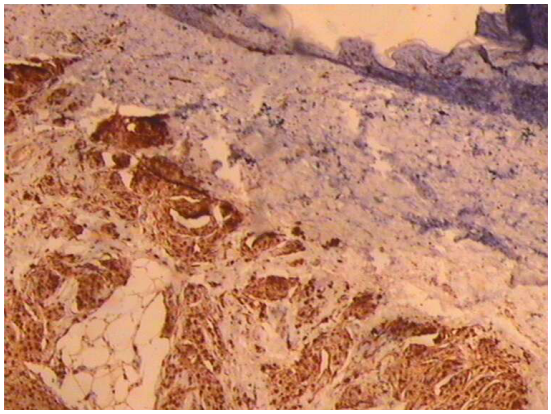

Figure 3: S100 protein staining of neurotized melanocytic nevus with a coarse, globoid, pattern ,S100 staining pattern (original magnifications ×100).

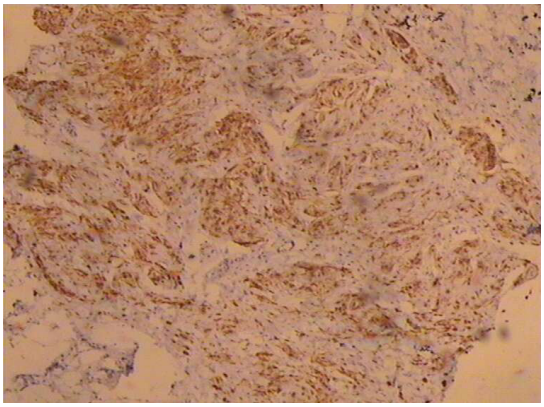

Figure 4: Neurotized melanocytic nevi with positive Melan-A staining of different neurotized areas, Most neurotized melanocytes show strong Melan-A staining. (original magnifications×100).

The clinical and histological differentiation between neurofibromas and neurotized melanocytic nevi may be difficult. The shape and rank pattern and location of cells, the appearance of specific cells,positive or negative Melan-A staining and S100 protein staining can distinguish neurofibromas and neurotized melanocytic nevi correctly and validly [5]. Her material with informed consent were obtained and approved by the ethics committee of the Second Affiliated Hospital of Harbin Medical University and family member signed informed consent.

Discussion

The timing and choice of treatment of GCNN continues to evolve under the influence of changing opinions regarding the purpose that minimizes the risk of malignant degeneration and at the same time provides optimal functional and aesthetic outcomes for the affected patients. Melanoma can arise in the first few years of life and excision should be considered as soon as possible. Choices for coverage include skin grafts, local flaps, expanded transposition flaps, expanded pedicle flaps, and free tissue transfer. Now the cultured skin autograft are sometimes used as a reconstruction of the large excised melanoma. Advances in tissue engineering, gene therapy, and other nonsurgical modalities e.g. dermabrasion, laser, chemical peels and so on, may offer patients an even better outcome than what can be achieved today.

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Competing interests

The authors declare that they have no competing interests.

Consent for publication

Not applicable.

Conflict of interest statement

There is no conflict of interest among the authors of this study and no objection to the selection and order of the authors.

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