Cutaneous blue nevi: Classification and malignant degeneration

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MM Al-Qattan, MF Stranc, BJ Anderson. Cutaneous blue nevi: classification and malignant degeneration. Can J Plast Surg 1994;2(3):130-132. A case of malignant melanoma arising in a congenital giant blue nevus on the arm of a 72-year-old male is reported. The literature is reviewed and a classification of cutaneous blue nevi presented. Malignant degeneration of different types of blue nevi is also discussed.

Key Words: *Blue nevus, Congenital giant nevus, Malignant melanoma*

Naevus bleu – classification et dégénérescence cancéreuse

RÉSUMÉ : Un cas de mélanome malin provenant d'un naevus bleu géant congénital au niveau du bras chez un homme de 72 ans est présenté ici. La littérature est passée en revue et une classification des naevi bleus est présentée. La dégénérescence cancéreuse de différents types de naevi bleus est également abordée.

Cutaneous blue nevi were first described by Tieche (1) who was a student of Jadassohn; the blue nevus is therefore also known as Jadassohn-Tieche nevus.

The nevomelanocytic cell is derived from the neural crest and migrates with peripheral nerves to the skin. By the second half of gestation there are no melanocytes seen in the dermis except for a few residual areas in the sacral region, the dorsal aspect of hands and feet, and the scalp. Blue nevi are thought to result from incomplete migration of melanocytes to the epidermis. Hence, blue nevi are most commonly seen in these residual areas (2).

The blue nevus cells are mainly found in the deep dermis and therefore appear clinically blue due to the Tyndall light-scattering effect of the overlying tissue.

A 72-year-old male presented with a large blue nevus of the left arm. The lesion was congenital but the patient recently noticed subcutaneous masses within the lesion.

Examination showed a 10xl0 cm blue nevus of the left lower arm with multiple adjacent satellite blue nevi (Figure 1). Multiple enlarged mobile subcutaneous nodules within the lesion were felt as well as ipsilateral enlarged axillary lymph nodes. A target blue nevus (a blue nevus with a depigmented halo at the periphery) was also noticed in the midline of the upper back. A skin biopsy of the arm lesion including a subcutaneous nodule showed malignant melanoma in a cellular blue nevus; a lymph node biopsy (Figure 2) confirmed malignant regional metastases.

There was no evidence of systemic metastases at the time of the initial presentation. However, the patient refused any treatment and died six months later with widespread brain, lung and liver metastases.

DISCUSSION

Classification of cutaneous blue nevi

Lever and Schaumburg-Lever (3) classified cutaneous blue nevi into four types: the common blue nevus, the cellular blue nevus, the combined blue nevus and the malignant blue nevus. Maize and Ackerman (2) prefer to classify blue nevi according to the morphologic features of the cells that compose them.

Cutaneous blue nevi can behave as benign, premalignant, locally malignant or malignant lesions and the following types have been described in the literature.

The common blue nevus (4,5) occurs as a small (less than 1 cm) well circumscribed blue lesion and is commonly found on the dorsal aspect of hands and feet. Histologically, bipolar melanocytes with dendritic prolongations lie grouped in irregular bundles in the dermis with fine melanin granules filling the cells and their processes. Except for one case report

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Figure 1) *A congenital plaque blue nevus of the left arm with malignant transformation in a 71-year-old patient. Note the halo blue nevus of the midback*

Figure 2) *Cross section of an enlarged axillary lymph node with regional metastases*

in which malignant transformation occurred at the age of 66 years (6), malignant melanomas are not known to arise from common blue nevi.

The benign cellular blue nevus (7,8) usually appears as a bluish nodule 1 to 5 cm in diameter with a smooth or irregular surface and is most commonly found over the sacral area and scalp. The most characteristic histological picture is the biphasic pattern. Highly cellular islands composed of scarcely pigmented spindle cells are seen alternating with more sclerotic areas with deeply pigmented bipolar dendritic melanocytes. Malignant degeneration of benign cellular blue nevi is rare but has been reported (9).

The target (Cockade or halo) blue nevus (10,11) is a cellular or common blue nevus that has lost colour from its periphery.

The giant plaque (eruptive or agminated) blue nevus is greater than 5 cm in diameter with multiple adjacent satellite lesions. Histologically, the lesions are either common or cellular blue nevi. Reviewing the literature, we found seven cases of plaque blue nevi. Six were benign (12-16), while the case reported by Hartmann et al (17) was a congenital plaque cellular blue nevus with malignant degeneration at the age of 41 years. In our case, malignancy occurred at the age of 72 years.

The combined blue nevus (18) is a blue nevus with an overlying melanocytic nevus, which may be junctional, compound, intradermal or rarely a spitz nevus. The combined blue nevus is a histologic rather than a clinical diagnosis.

Pseudometastasizing pseudomelanoma is a cellular blue nevus with benign metastasis. The finding of thyroidal and decidual tissues in the draining nodes, with no evidence of malignancy, has given rise to the concept of 'benign metastasis' (19). In a few benign cellular blue nevi, small groups of well differentiated nevus cells are found in the marginal sinuses of the lymph nodes, with no invasion. It can be assumed that these cells do not represent true metastases, but were passively transported to the lymph nodes. Following excision of the primary lesions and involved nodes, the reported cases remained asymptomatic on long term follow-up (20).

Locally invasive congenital cellular blue nevi of the scalp have two typical clinical presentations. The first is a locally invasive scalp nevus with no evidence of malignancy at birth, but failure to carry out complete excision results in local recurrence, malignant degeneration and eventually distant metastases (21,22). The second presentation is a cellular blue nevus of the scalp with focal areas of malignancy seen at birth. There is typically an underlying skull defect with invasion of the dura (23).

The malignant blue nevus (24) is a rare tumour arising in a blue nevus or nevus of ota or it may be malignant from the start. The clinical picture, metastases and prognosis of malignant blue nevi are similar to malignant melanoma and should be treated as such.

Malignant degeneration of blue nevi

From the above classification it becomes evident that the cutaneous blue nevus is a complex entity. It can behave as a benign, premalignant, locally malignant or malignant lesion. The recognition and understanding of these lesions will help to plan an appropriate management.

It is important to differentiate between blue nevi and other melanocytic nevi. The risk of malignancy in melanocytic nevi is the subject of considerable debate but it is generally agreed that there is a higher risk in dysplastic nevi, Hutchinson's freckles and congenital giant hairy nevi (25). There is also a general agreement that the probable risk of malignancy in congenital giant hairy nevus is higher during childhood than it is in adult life (26-28).

On the other hand, both the malignant potential of benign blue nevi and the age at which malignant transformation occurs will vary according to the type of blue nevus. The most aggressive type is the cellular blue nevus of the scalp and invasion/malignant transformation tends to occur soon after birth or in early childhood (21,22). The probable risk of malignancy of giant plaque blue nevi is higher than cellular and common blue nevi and malignant degeneration of blue nevi outside the scalp region tends to occur in adulthood rather than childhood (6,17,24).

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