Infiltrating Giant Congenital Cellular Blue Nevus Of Neck Presenting As Melanoma

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Citation

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Abstract

Blue nevus is an uncommon pigmented lesion of dermal melanocytes. By convention, two well defined histologic variants, designated as "common" and "cellular", have been recognized. These lesions have attracted much attention due to its confusion with malignant melanoma. We present a case of a giant congenital cellular blue nevus of neck clinically presenting as malignant melanoma and infiltrating vessels and underlying muscles resulting in incomplete removal. It is important to differentiate this lesion from malignant blue nevus and malignant melanoma on the basis of presence or absence of severe nuclear pleomorphism, nucleoli, mitotic activity and necrosis. However, malignant transformation is known to occur in these lesions and the importance of careful follow up is mandatory. This case highlights the difficulty in diagnosing this lesion and emphasizes the need for long term follow up in view of its uncertain malignant potential.

INTRODUCTION

Blue nevi are common lesions which represent arrested melanocytic migration. Histologically, there is presence of pigmented spindle and dendritic melanocytes in a focal area of reticular dermis. Cellular blue nevus is a distinctive variant of blue nevus suspected of being malignant because of its large size and intense pigmentation. These lesions tend to be 1-3 cm in diameter, usually solitary and present as elevated smooth-surfaced gray-blue papules or plaques. They are most commonly seen on buttocks, sacral region, occasionally on dorsal aspect of hands and feet and rarely on head and neck. Blue nevi can develop at any age but are usually noticed in second decade of life or later and are twice as common in women as in men. Although rare, malignant degeneration of cellular blue nevi can occur.

CASE REPORT

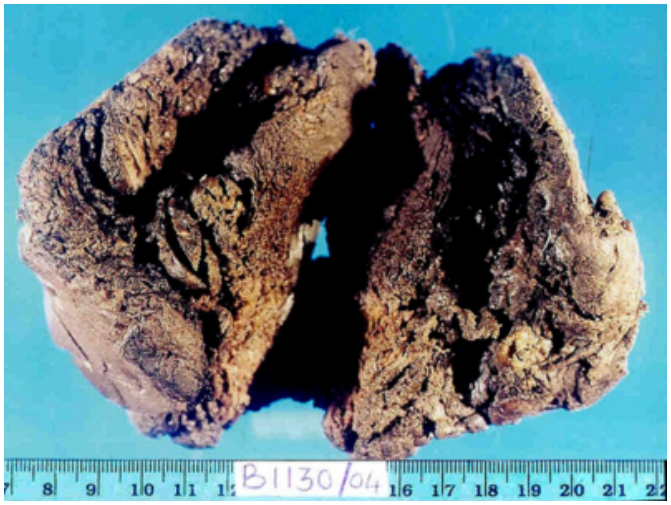
We report a case of an unusually large pigmented tumor present in the neck of a 20yr old man extending from behind the left ear to the left angle of mandible and infiltrating left sternocledomastoid muscle (SCM). The tumor began as a small nodule behind the ear, a few weeks after birth, and gradually increased in size over the years. Patient was otherwise asymptomatic. A diagnosis of fibromatosis was made on the basis of FNAC report from an outside center and surgical removal was planned for cosmetic reasons. Intraoperatively, the tumor and surrounding vessels, muscles and facial nerve were all seen to be pigmented and tumor was seen infiltrating deep into SCM. Complete surgical resection was not possible because of infiltration and encasement of vital structures like facial nerve and carotid artery. A small skin flap was excised along with the tumor and the defect repaired. Postoperative period was uneventful.

HISTOPATHOLOGICAL FEATURES

Macroscopic examination of specimen submitted for histopathological examination showed a black tumor mass measuring 15x10x5 cm with a small skin flap (Fig 1).

Figure 1

Figure 1: Skin covered excised irregular tumor mass with pigmented cut surface



Microscopy revealed an atrophic epidermis with grenz zone and well-circumscribed nests of tumor cells, exhibiting a biphasic pattern of pigmented dendritic and non-pigmented spindle cells in reticular dermis (Fig 2), along skin adnexae (Fig 3) and extending deep into subcutis and skeletal muscle (Fig 4). Pigment in dendritic cells was demonstrated as melanin by Schmorl's reaction and Masson's Fontana stain (Fig 4, inset). There was no evidence of mitosis or necrosis and regional lymph nodes were free of any tumor deposits. After melanin bleach cells were seen as uniform and bland. With this picture diagnosis of malignant melanoma was ruled out and a provisional diagnosis of cellular blue nevus was made. The size of the lesion, history of presence since birth and permeation along skin adenexa were correlated with the histological picture and the final diagnosis of Giant congenital nevus was made.

Figure 2

Figure 2: Biphasic pattern of pigmented and non-pigmented tumor cells.

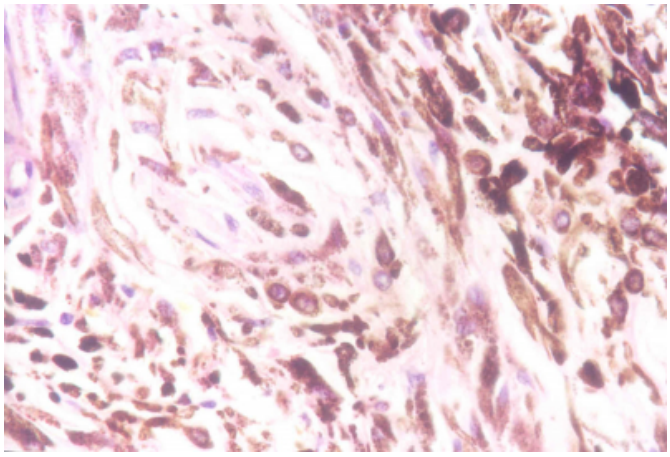


Figure 3

Figure 3: Tumor cells in bundles permeating along hair follicle.

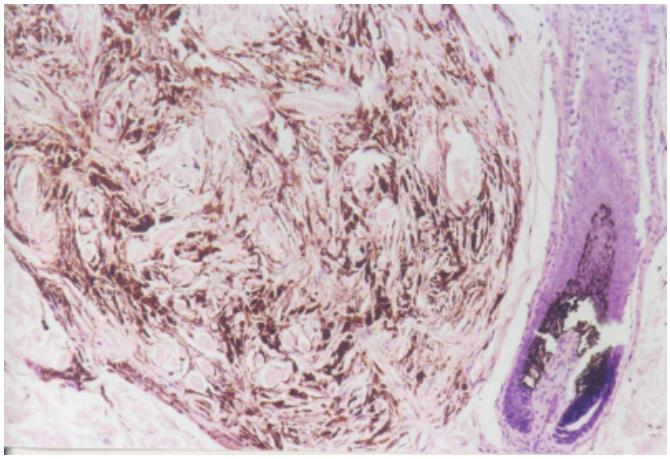
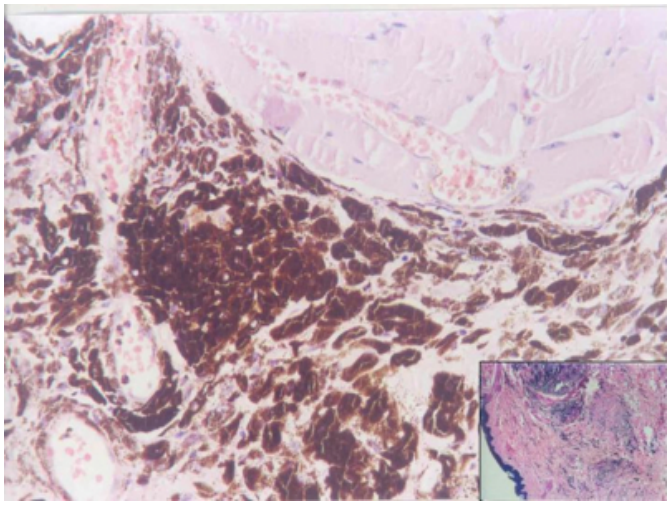


Figure 4

Figure 4: Tumor cells infiltrating skeletal muscles with inset showing pigment by Massons Fontana stain.



DISCUSSION

Giant congenital nevi differ from acquired nevi in histological appearance and are more complex than the non giant congenital nevi, while cellular blue nevus maybe found as a small component of giant pigmented nevus, occasional cases wherein the entire scalp lesion consisted of cellular blue nevus have been reported. ([1]) A common form of congenital nevomelanocytic nevus (CNN) or giant pigmented nevus. Congenital nevi differ from the more common acquired variety because of their generally larger size, tendency to involve reticular dermis and subcutaneous tissue, single cell permeation of dermal collagen bundles and involvement of skin adenexa, muscles nerves and vessels. ([2,3]) Majority of congenital nevi are smaller than 3-4 mm in diameter, while very large congenital nevi are present in 1 in 20,000 to 1 in 50,000 newborns. Congenital nevi are apparent at birth and may be found in up to one percent of the population. They usually appear for the first time between 1 month and 2 yrs of life. While most CNN are small and singular, there is no completely satisfactory way to classify them. Giant has been generally defined as a lesion as large as the patients palm for the face or neck, and twice this area for other anatomic sites. ([4]) Other authors have divided CNN according to the largest diameter (Small as <1.5cm, Medium as 1.5 to 19.9 cm and Large >= 20 cm)

The histology of giant CNN, maybe divided into nevus cell, neuroid type, epitheleoid and/ or spindle cell, dermal melanocytic or mixed type. In nevus cell type nevomelanocytes are seen in epidermis as well as papillary dermis as sheets, nests and cords. In case of neuroid type of giant CNN formations such as neuroid tubes and corpuscles are present. In the epitheleoid and / or spindle cell type of giant CNN, the dermis maybe infiltrated in whole or in part by nests and sheets of epitheloid and / or spindle cells, often intermixing with neuroid elements and ordinary nevomelanocytes. In the dermal melanocytic type the appearance maybe that of a giant blue nevus or the lesion may have elements of blue nevus (common and cellular). Heavily pigmented spindle-shaped melanocytes may occur alone or intermixed with nevomelanocytes with long occasional branching dendritic processes seen lying as irregular bundles in the dermis. In cellular blue nevus there is usually a component of common blue nevus plus fascicles of spindle-shaped cells with ovoid nuclei and abundant pale cytoplasm with little or no melanin.

Giant congenital blue nevi have been associated with development of melanoma and cases of spread to lymph nodes and death due to local invasion and visceral metastasis have been reported. ([5]) The lifetime risk of melanoma for patients with very large / giant CNN has been estimated to be at least 6.3%. ([6]) Aloi F, etal ([7]) reported 6 cases of malignant transformation of preexisting cellular blue nevus. The lesions were present since childhood and had rapidly enlarged. Some of them were heavily pigmented and some showed evidence of mitosis and necrosis. Cases of melanoma mimicking cellular blue nevus have been reported. But in all cases malignancy was evidenced by increased mitotic rate, necrosis, nuclear atypia, pleomorphism and prominent nucleoli. ([8]) Granter SR reviewed clinicopathological features of 10 cases of malignant blue nevi, 6 cases proved to be de nevo melanoma mimicking cellular blue nevus; two arose in association with common blue nevus and two with cellular blue nevus. Head and neck area was found to be the most common location and the patient's age ranged from 11-77 yrs. ([8])

While there is disagreement about the exact frequency of association between CNN and melanoma, risk of melanoma arising in large melanocytic nevi has been reported. Therefore excision should be considered as early as possible and long term follow-up advised after resection. ([9]) However the enormous size and infiltrative pattern of large CNN do not allow complete removal and treatment goal is to remove as much as possible while preserving functions and improving cosmetic appearance. In such cases photographic follow up of these lesions maybe the most effective means of reducing the morbidity and mortality of cutaneous melanoma.

In our case, after a clinicopathological correlation a diagnosis of giant CNN was made. Surgery was performed almost 20 yrs after occurrence of the nevus and during the surgery it was impossible to remove all the tumor tissue because of the involvement of vital structures. As the lifetime risk of melanoma for patients with very large CNN has been estimated to be at least 6.3 % and because of the difficulty found incomplete resection of the tumor, the prognosis of our patient appears to be poor.

The patient and his family were counseled about the risk of malignancy and encouraged to perform regular self-skin examination of the skin to look for any change in colour/surface at site of surgical incision or sudden appearance of any nodule or any ulceration, which should be reported to the surgeon. A regular visit to the specialist clinics has been advised to have baseline colour photographs taken. Whole body and close-up photographs may enable early detection of change in existing naevi, especially where a trained specialist evaluates dermoscopic views of individual lesions. Possible review in 3-6 months by specialist/surgeon and opportunistic skin checks by general practitioner (ie when visiting practitioner for other reasons) were also advised. At the time of communicating this report the patient was on post-operative follow-up for 12 months and did not show any evidence of recurrence or pressure symptoms.

This case report stresses the importance of regular reviews post-operatively for early detection of any malignant change.

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