

Congenital Black Hairy Nevus

Jaswir Singh, Kuldish, Sanjeev

Department of Pediatrics, Government Medical College, Patiala, Punjab, India

Abstract: Black pigmented hairy nevi are congenital melanocytic nevi. Congenital nevi are present in approximately 2-3% of neonates. A four years old male child presented with hairy nevus on the left side of forehead adjoining the left eye brow present since birth, 5cm × 6cm, progressive and remained asymptomatic. Such nevus is of great cosmetic value and needs surgical intervention.

INTRODUCTION

Melanocytic nevus of the face is a unique form of congenital nevus, fortunately rare and typically hair bearing. These lesions have a relatively high risk of becoming malignant. They develop probably between 40 days of gestation and six months in utero. It develops from neuro ectodermal cells between the 8th to 24th weeks of pregnancy. Body protein HGE/SF (hepatocyte growth factors/scatters factors) seems to be responsible for encouraging these neuroectodermal cells to develop, migrate and scatter. In those of us with nevus, it seems that we have too much of the wrong type body protein. HGE/SF in some cells, so we develop extra pigment and abnormal skin cells called nevus cells. Congenital nevi are generally seen in 1 in every 20,000 children. Whereas the giant variety involving much of the body surface area are less common, possibly around one in every 200,000. They are often round or oval, clearly demarcated and sometimes slightly intact. Pigmentation is usual, although some congenital nevi have a speckled appearance. Coarse dark hair may be present. The congenital nevi occur in 1-1.25% of neonates and with passage of time, may change from flat, pale, tan macule to elevated verrucous hairy lesions. The head and neck area involve approximately 15% of all nevus. Intraoral occurrence is extremely rare. Congenital nevi are significant because of their much higher incidence of melanoma transformation. These lesions suggests may be genetically inherited. They may be associated with put birth defects. Lifetime risk for malignant degeneration in a large congenital nevus is approximately 6%. People with nevus presenting neurological symptoms, like worsening headache and vomiting, need to have an MRI of the brain. Those with seizures also need to have repeated MRI performed periodically. Staged surgical excision remains the mainstay of treatment for large congenital melanocytic nevi resurfacing with skin graft or tissue expanders and flaps. Histologically, nevi are transformed melanocytes, which are normally highly dendritic cells interspersed among basal keratinocytes. Finding of a culture of melanocytes from such lesion showed chromosome rearrangement involving 1p, 12q and 19p. The giant nevi might be associated with several diseases; neurocutaneous melanosis, diffuse lipomatosis, structural brain malformation, hypertrophy of skull bones, limb atrophy, skeletal asymmetry involving both hyper and hypoplasia, von Recklinghausen disease and vitiligo.

CASE REPORT

Four years old male child presented in pediatric OPD department with complain of pain abdomen for the last 15 days. On examination patient was anemic, had black colored patch on the left side of forehead adjoining the left eye brow measuring about 5cm× 6cm. This black patch was hairy, no itching, no discharge, painless and with normal temperature. Heart rate was 110/min; respiratory rate was 26/min, blood pressure 100/70mmHg, no hepatosplenomegaly or lymphadenopathy.

Laboratory investigation shows Hemoglobin-9gm% ; TLC-10500/cumm; DLC-Neutrophils 55, lymphocytes 38, eosinophil 07; Stool examination and urine culture was normal, PBF was microcytic hypochromic. Patient was treated with deworming drugs and hematonic in pediatric OPD. Skin opinion was taken from dermatological department and confirmed, it was a case of congenital big black hairy nevus.

DISCUSSION

Congenital hairy nevus is a hyper pigmented lesion that has its color from the



melanin pigment of nevomelanocytes. Nevomelanocytes, derivatives of melanoblasts, compose the cellular format of the neoplasm¹. Multiple definitions have been used to classify nevi into small, medium, or giant. These include diameter size, total body surface area (TBSA), and ability to excise in one surgical setting². Based on diameter, CNN are characterized as small (<1.5 cm), medium (1.5-19.5 cm), and large or giant (>20 cm in adolescents and adults or predicted to reach 20cm by adulthood). Giant hairy nevi on the scalp and neck may be associated with leptomeningeal melanocytosis and neurologic disorders that include neurofibromatosis, epilepsy or focal neurologic abnormalities. Lesions over the vertebral column may be associated with spina bifida or meningomyelocele. Infants with lesions on the scalp or posterior midline should undergo neuroimaging studies to detect associated conditions that may affect treatment and prognosis³. CNN are present at birth or soon thereafter. Delay in appearance of surface pigmentation may occur from age 1 month to 2 years in the rare "tardive" type. The incidence for small nevi is 1 in 100 births; for a CNN larger than 9.9 cm in diameter medium nevi, 6 in 1000 births; and for large nevi, 1 in 20,000 births⁴ in 1 per 20,000 newborns; a CNN larger than 20 cm in diameter occur in 1 per 500,000 newborns⁵. Prophylactic excision of all giant and large hairy nevi is indicated⁶. In 60% of these cases malignancies can develop in early childhood⁷. In one series, 10% to 20% malignant melanomas arising in congenital nevi were diagnosed before three years of age⁸. In addition, concerns about lack of parameters to predict as to which nevi are susceptible to malignant transformation, compounded with the fact that the diagnosis gets delayed and the course in case of a malignant change is invariably fatal are reasonable⁹. A recent review has concluded that an early aggressive approach to these lesions is responsible for the low risk of malignant melanoma reported in various series^{9,10}. Surgical excision results in the debulking of tissue at risk for the development of malignant melanoma. Tissue expansion forms the most widely used modality of treatment^{11,12}. Timely use of these modalities can minimize the risks of malignancy and reducing the psychological stress on the parents and the child.

REFERENCES

1. Rhodes AR. Benign neoplasias and hyperplasias of melanocytes. In: Fitzpatrick's Dermatology in General Medicine Year. 5th ed. 1999:1026-1032.
2. Nevi and Malignant Melanoma. In: Habif TP. Clinical Dermatology: A Color Guide to Diagnosis and Therapy. 4th. Edinburgh: Mosby; 2004:776-77.
3. Reed WB, Becker SW Sr, Becker SW Jr, Nickel WR. "Giant pigmented" nevi, melanomas and leptomeningeal melanocytosis. Arch Dermatol 1965;91:101-119.
4. Rhodes AR. Melanocytic precursors of cutaneous melanoma. Estimated risks and guidelines for management. Med Clin North Am 1986; 70(1):3-37.
5. Rhodes AR, Weinstein MA, Fitzpatrick TB, et al. Risk factors for cutaneous melanoma: a practical method of recognizing predisposed individuals. JAMA. 1987; 258:3146-3154.
6. Dellon AL, Edelson RL, Chretien PB. Defining the malignant potential of the giant pigmented nevus. Plast Reconstr Surg. 1976;57:611-17.
7. Rhodes AR, Wood WC, Sober AJ, Mihm MC. Nonepidermal origin of malignant melanoma associated with giant congenital nevocellular nevus. Plast Reconstr Surg. 1981;67:782-90.
8. Kaplan EN. The risk of malignancy in large congenital nevi. Plast Reconstr Surg. 1974;53:421-28.
9. Huemer GM. The value of full-thickness skin grafts in reconstruction of the periorbital region. Plast Reconstr Surg. 2008;121:1857-58.
10. Bruce S, Bauer MD, Frank A, Vicari MD. An approach to excision of congenital giant pigmented nevi in infancy and early childhood. Plast Reconstr Surg. 1988;82:1012-21.
11. Tannous ZS, Mihm MC, Jr, Sober AJ, Duncan LM. Congenital melanocytic nevi: Clinical and histopathologic features, risk of melanoma, and clinical management. J Am Acad Dermatol. 2005;52:197-203.
12. MacKie RM, English J, Aitchison TC, Fitzsimons CP, Wilson P. The number and distribution of benign pigmented moles (melanocytic naevi) in a healthy British population. Br J Dermatol. 1985;13:167-74.

Correspondence: Dr Jaswir Singh, #5084, Urban Estate Phase I, 26 ACRE-C, ITBP Wall, Patiala PB 147002, Tel.: 0175 2281088, Mobile: 9815664208