Giant Congenital Hairy Nevus On The Scalp
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Abstract

Congenital nevi or birth marks are hyperpigmented macular lesions that are derivatives of the melanoblasts. They occur in less than 1% of the neonates in any site of the body. Nevi larger than 9.9 cm in diameter occur in 1 per 20,000 newborns. 50% have hair on them. They vary in size, shape, color and texture. They have the propensity to become malignant as the child grows, so, regular follow up should be done to interfere if there is a risk of malignancy. The risk depends mainly on the size of the nevus. We report here a giant hairy nevus on the scalp that have a size of about 15cm and occupied almost half of the scalp surface. no other associated anomalies were found & the patient is under regular follow up in our clinic.

Key words: nevus, hairy, size, malignancy.

Case

This is a newborn male baby who was delivered at term by normal vaginal delivery and was found to have hair on one side of the scalp. In the first instance, one would think that he is bald in one side but at closer views, a blackish area was seen underneath the area of the hair. We found a big oval shaped hairy nevus on just one side of the scalp with a diameter of 15cm. the margins were well defined and the hair was thick.

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Fig 2

There was short thin hair on the other side of the scalp with normal color of the skin beneath it. His photos are shown below.
No other lesions were noticed on the rest of the body. The rest of the examination was normal.
There was a negative family history of similar lesions.
The baby was discharged home the next day and he is under regular follow up in our clinic

Discussion

The congenital nevomelanocytic nevus (CNN), and known commonly as the congenital hairy nevus is a hyperpigmented lesion that has it's color from the melanin pigment of nevomelanocytes. Nevomelanocytes, derivatives of melanoblasts, compose the cellular format of the neoplasm.

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 setting.

Nevi vary greatly in size, shape, surface texture, and hairiness. The color can vary from light tan to nearly black. Most are shades of brown. Some have fine downy hair; many have long, thick, darker, coarser or curlier hair. Coarse surface hairs develop in more than 50% of lesions and may occur during the first 1-2 years of life. Cutting or shaving this hair will not increase its growth.
The congenital nevi that are small to medium typically appear as tan or brown irregularly shaped macules. Overtime, the nevi may become elevated and intensely pigmented, sometimes covered with coarse dark hair. Small to medium nevi frequently darken at puberty, with pregnancy, or with the use of oral contraceptives. Giant congenital nevi or giant hairy nevi have an irregular margin often with a verrucous surface. Their color is typically dark brown to black and 95% of them have dark, coarse surface hair. Satellite lesions are often present beyond the periphery of the main lesion and may be scattered over the entire skin surface. Around the age of 10 the giant nevus becomes more elevated, verrucous, and hyperkeratotic and the surface hair thicker.

A giant nevus often lacks subcutaneous fat. Because of this, an infant or an adult may look "thinner" over the area of the nevus. Very rarely a nevus child will actually have less flesh underneath the nevus, with atrophy of the arm or leg, so the area looks shrunken. They may also look slightly misshapen. Smaller nevi often appear inside a larger nevus. Different nevi penetrate the skin to different depths. If the nevus is thin, the skin is also thin, fragile, and easily "pinched." If the nevus is thick, the skin may be lumpy or folded or bulbous-looking. Occasionally some people will have soft-tissue tumors underneath the nevus, often lipomas or neurofibromas, giving the skin a very bumpy, folded appearance. Nevus skin is easily damaged and doesn't seem to heal as well as normal skin.

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 meningomyelecele. Infants with lesions on the scalp or posterior midline should undergo neuroimaging studies to detect associated conditions that may affect treatment and prognosis. 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 medium nevi, 6 in 1000 births; and for large nevi, 1 in 20,000 births. A CNN larger than 9.9 cm in diameter occurs in 1 per 20,000 newborns; a CNN larger than 20 cm in diameter occur in 1 per 500,000 newborns. An equal prevalence exists in males and females.

Autosomal dominant inheritance with incomplete penetrance or multifactorial determination occurs in families with small CNN. CNN appear in all races, but, paradoxically, the frequency of small CNN is slightly higher in some populations such as blacks who are at lower risk of developing melanoma than white.

The cause of congenital moles may have recently been discovered. Researchers have known that the skin and nerves of a fetus develop from the same primary cells of the body, which are called neuroectodermal cells, between the 8th and 24th week of pregnancy. Melanoblasts migrate from the neural crest between weeks 8 and 10 of gestation. CNN develop in utero after the melanocytes appear but before the sixth antenatal month. Researchers think that a body protein called HGF/SF (hepatocyte growth factor/scatter factor) seems to be responsible for encouraging these neuroectodermal cells to develop, migrate, and "scatter." In those of us with a nevus, it seems that we have too much/wrong type of this body protein HGF/SF in some, not all, of our cells, so we develop extra pigment and abnormal skin cells called nevus cells. These cells "scatter" around, so we have nevi "scattered" all over us. If we have lots of nevus cells scattered on the skin, there seems to be a good chance that we have the same cells scattered in the brain. That may be why some of us develop nerve symptoms like hydrocephalus, melanoma, and neurocutaneous melanosis.

Congenital nevi usually form in the fetus between 2nd and 6th month of pregnancy. A nevus is often symmetrical around the spinal cord because the spinal cord and skin develop together from those same early neuroectodermal cells. A very few cases of large nevi appearing after birth, called tardive nevi, are known. CNN are most common in Caucasians and least common in Africans, with intermediate frequency in Hispanics and Asians. It seems that the darker the normal skin, the less is the risk of a giant nevus. Several instances of identical twins, one with a large mole and the other without, are known. Identical twins do not share the same HGF/SF.

The potential for large congenital nevi to become malignant is significant and is an important consideration in the treatment and management of this entity. The size of the lesion correlates with the potential for malignant transformation. Malignancy should be suspected with focal growth, pain, bleeding, ulceration, significant pigmenary change, or pruritus. The risk of malignant melanoma in patients with small to medium ones range from 2.6% to 4.9%, while for giant nevi the risk is felt to be approximately 6%. Small congenital nevi pose a much greater practical risk since they occur more frequently than giant nevi. Very large congenital nevi account for less than 0.1% of cutaneous melanomas, whereas small varieties of congenital nevi may account for 15% of cutaneous melanomas.

Malignant change of small nevi usually occurs after puberty, whereas 60% of all malignant melanomas arising from giant nevi will develop in the first decade of life. In contrast to the greater frequency of congenital nevi in blacks, the frequency of melanoma in
blacks is lower than in whites by a factor of 10 to 20\(^3\). The management and treatment of patients with CNN remains controversial and depends on the lesion's size, location, and propensity for malignant transformation\(^6-11\). No absolute guidelines can be recommended. Management is individualized and centered around: cosmetic appearance, risk of neoplastic proliferation, and the psychological impact on the patient and family...

Aesthetic considerations are important. Surgical treatment of giant or large CNN is addressed at age 6 months. Procedures used in surgical treatment include serial excision and reconstruction with skin grafting, tissue expansion, local rotation flaps, and free tissue transfer. Adjunctive treatment options include chemical peels, dermabrasion, and laser surgeries. Surgical excision remains the mainstay of treatment, since other adjunctive treatment options do not fully eradicate the nevus cells. Management of small lesions includes close monitoring with photographic documentation versus surgical excision. Because it is impractical to prophylactically excise all non–giant congenital nevi, yearly examination for the first 3 years of life is recommended, with reassessment every 2- to 5 years afterwards depending on the confidence of the parents to monitor the lesions\(^6\). Biopsy specimens are obtained from lesions that undergo suspicious alteration. The impact of giant hairy nevi is greater because of the considerable cosmetic disfigurement as well as the higher malignant potential. It is more difficult to recognize early malignant changes within giant pigmented nevi because of their typical irregular surface and pigmentation. Some have recommended total surgical excision as early as technically practical, while others are more cautious because of the large areas involved and the resulting disfigurement and complications from scarring. Due to the depth of some lesions, especially if the leptomeninges are involved, excision may not eliminate the risk for developing melanoma\(^11\).

When a large congenital nevus involves the head and neck or midline over the trunk, associated meningeal melanocytosis may be observed, occasionally complicated by seizures, focal neurologic defects, obstructive hydrocephalus, or malignant changes. Radiographic imaging, including MRI, is warranted to evaluate melanocytic depositions in the CNS. The baseline MRI should be obtained when the patient is aged 4-6 months. Serial MRIs are frequently required in patients with meningeal melanocytosis\(^6\).

References
