Giant Pigmented Hairy Nevus

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The pigmented hairy nevus that covers the entire right upper arm of this 18-month-old boy has been present since birth. More than 95% of large, congenital pigmented nevi have a hairy component.

The incidence of congenital pigmented nevi varies with the lesion's size; nevi less than 1.5 cm in diameter occur in 1% of all newborns, whereas lesions greater than 20 cm in diameter are found in 1 in 20,000 neonates. Depending on the lesion's size, the patient's lifetime risk of malignant melanoma ranges between 2.6% and 6.3%.

Giant pigmented hairy nevi of the scalp and neck may be associated with leptomeningeal melanocytosis and neurologic disorders such as epilepsy or other focal neurologic abnormalities. Those nevi that overlay the vertebral column can be associated with spina bifida or meningomyelocele.

The appropriate age for removal of a benign-appearing nevus is controversial, since malignant degeneration of small to medium congenital pigmented nevi usually does not occur until after puberty. Nevi that demonstrate suspicious changes need to be excised without regard to the patient's age.

Treatment of large nevi, such as seen in this patient, presents a difficult surgical dilemma.

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