ANSWERS TO MEDICAL QUIZ

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A ١. Extensive pigmental macules involving chest, trunk and extremities.

A ٢. The most likely diagnosis is congenital giant melanocytic nevus.

A ٣. Congenital melanocytic nevi are pigmented macules or plaques that are present at birth or early infancy in approximately 1% of children. The lesions are often tan at birth and become darker and hairier during infancy. Congenital nevi can be divided into small (< 2 cm), medium (2 to 10 cm) and giant (> 10 cm) lesions. Most nevi are small to medium in size, with giant congenital nevi occurring in only 1 in 20,000 newborns[1,2].

The malignant potential of congenital nevi remains in area of great controversy. Risk of malignant transformation before 15 years of age is 1 in 10,000, and maximum risk increases to 1 in 3700 between the ages of 15[2] and 35 years in black patients. The risk of malignant transformation in general population for small and medium congenital nevi is unknown, and there are no universal guidelines for their management. Some experts advocate prophylactic excision of these nevi, whereas others advocate close observations of the nevi. Most dermatologists agree that removal of these nevi can wait until childhood, when local anesthesia and outpatient surgery are feasible.

The risk of malignant transformation of giant congenital nevi is another controversial issue and is reported to be between 2% and 15%, as compared to the general population's lifetime risk of melanoma of 1%. These large lesions warrant close observation and serial photography. Careful annual or semiannual examination with palpation of the nevi is essential, as melanoma can arise from deep portions of the nevi with little or no apparent surface alterations.

Pediatricians caring for infants or children with giant congenital nevi should also be aware of the potential for neurocutaneous melanosis; this is, the coexistence of large or multiple congenital melanocytic nevi and benign or malignant pigment cell tumors. The prevalence of this condition is unknown but appears to occur more commonly with congenital melanocytic nevi located on the scalp, face or neck. Signs and symptoms of neurocutaneous melanosis usually present during the first 2 years of life; however, they may become apparent during the second or third decades of life. Affected individuals may present with neurologic manifestations of increased intracranial pressure, spinal cord compression, or mass lesions. The development of leptomeningeal melanoma has a very poor prognosis and is usually fatal. Diagnostic procedures such as cerebrospinal fluid cytology and magnetic resonance imaging with gadolinium contrast should be considered in children with giant congenital nevi [3].

REFERENCES

1. Harwitz S. Clinical pediatric dermatology. Philadelphia: WB Saunders, 1993.

2. Shpall S, Frieden I, Chesney M, et al. Risk of malignant transformation of congenital melanocytic nevi in blacks. Pediatr Dermatol 1994;11:204-8.

3. Kadonaga JN, Freiden 1J. Neurocutaneous melanosis: Definition and review of the literature. J Am Acad Dermatol 1991;24:747-55.

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