

Congenital Nevus- A Rare Case Report



Medical Science

KEYWORDS : Nevus, Melanocytic, Melanoma, Neuroectoderm

Dr.K.R.Umadevi

Associate Prof, Dept of Pathology, Sri Muthukumaran Medical College Hospital and Research Institute, Chikkarayapuram, Chennai600069, Tamilnadu

Dr.Ezhilvizhi.A

Professor, Dept of Pathology, Sri Muthukumaran Medical College Hospital and Research Institute, Chikkarayapuram, Chennai600069, Tamilnadu

ABSTRACT

Congenital nevi are benign nevomelanocytic proliferations present at birth. Small congenital nevi are defined as those less than 1.5 cm in their largest diameter, medium-sized congenital nevi as those 1.5 to 19.9 cm, and large or giant congenital nevi as those greater or equal to 20 cm. 1% of the newborns have a biopsy-proven nevomelanocytic nevus. However, giant congenital nevi are rare with an incidence of approximately 1 in 20,000 births. Here we present 35 years old man with a swelling from childhood. Clinically, congenital nevi are round to oval in shape with regular, smooth, and well-demarcated border. Their clinical appearances may change with age. In our case swelling was round to oval with 2 satellite nodules. As the child grows, the nevi become progressively darker with uniform brown to dark brown or black color and they acquire long, coarse, darkly pigmented hairs. Occasionally, they may show considerable variation in pigment color within the background color, irregular border, and sometimes nodule formation. Such changes should be always evaluated for any malignant transformation. The lifetime risk of melanoma for patients with giant congenital nevi ranges from 5% to 10%. The risk is greatest between the ages of 5 and 10 years. The issue of malignant transformation of small congenital nevi is very controversial. Currently, there exists no consensus regarding their increased risk of developing melanoma or their management. However, many authors believe that small congenital nevi may be precursors for at least some cases of cutaneous melanoma but in our case no malignant changes were identified

Case Report

35 years old male presented with a swelling in the scalp to our Surgical OPD since Childhood. On examination the swelling was 10*8*4 cm, Lobulated with Satellite nodules Largest measuring 6*5*4 cm. Firm with ill defined margins. No H/o pain or itching. No H/o Neurological deficits. Not fluctuant. The size and the colour changes with the age. Diagnosis was made from Microscopic examination and Immunohistochemical markers.

X- ray - no calcification and no intra cranial extension. Clinically it was diagnosed as Neurolipoma.

Excision biopsy was done and sent to Histopathological Examination.

Grossly Skin covered Swelling measuring 8*6*2cm, External surface was Lobulated with many small satellite nodules larger one measuring 5*4*3 cm and smaller one measuring 0.5 cm. Cut surface is grayish white, firm, with ill defined margin.

Microscopy-Sections show thinned out Epidermis with Nevus cells arranged in Nests with increased melanin pigment Fig1 seen in Epidermo-dermal junction, Reticular Dermis and extending in to the Dermis, around appendages and up to the subcutaneous fat. Fig2, 3 Dermal component of Nevi show variable cellularity Fig 4 and evidence of Neural differentiation. Fig5. Showing S100 Protein Immunological marker positive demonstrating Neural differentiation. No evidence of Malignant melanoma is seen in the sections studied

All the Surgical margins are free of tumour

Impression :Picture is suggestive of Congenital Nevi- medium size

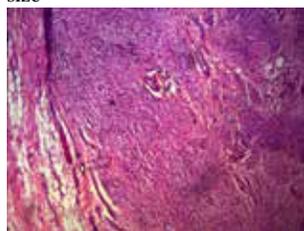


Fig – 1 Low power view of Congenital Nevus

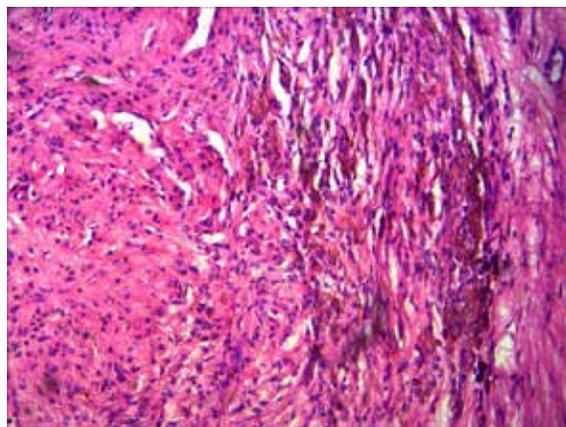


Fig – 2 Low power view of Congenital nevus

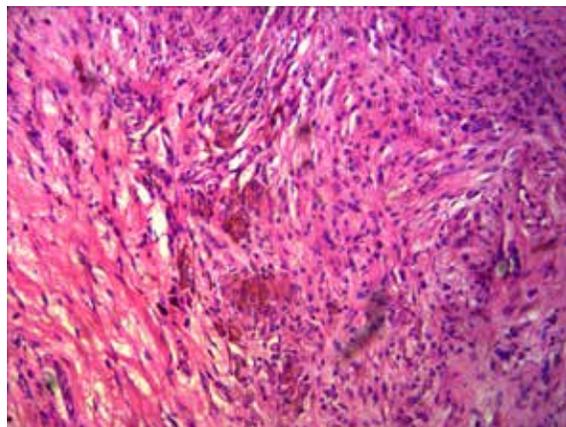


Fig – 3 High power view of Congenital nevus

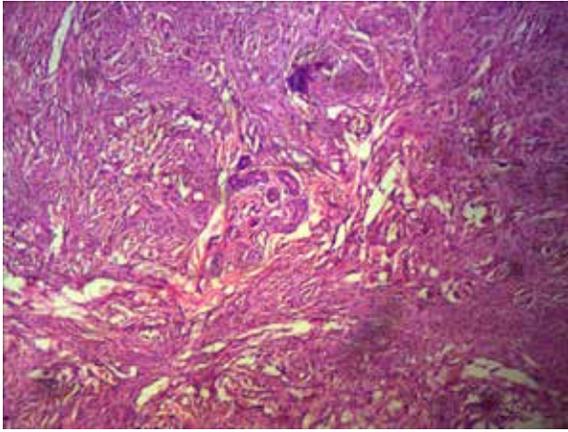


Fig - 4 Showing Neural Differentiation

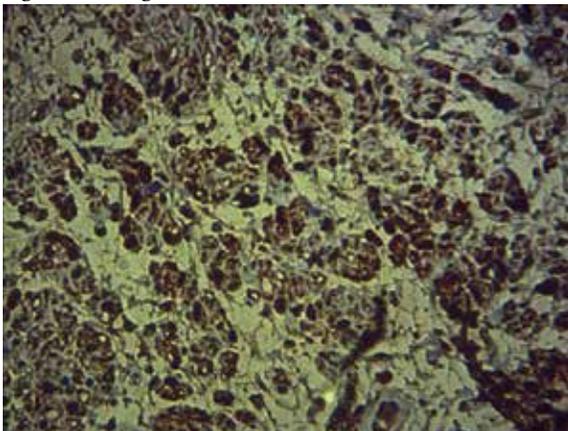


Fig - 5, Showing S100 protein Positivity

Clinical follow up for malignant melanoma is done since diagnosis for the past 3 months, so far no evidence of Malignant Melanoma.

Incidence

Congenital nevi are one of several known risk factors for the eventual development of Melanoma. Fortunately, melanoma remains an uncommon malignancy in prepubertal children, with an annual incidence of 0.7 cases per million children aged 0-9 years. Patient concerns regarding changing or worrisome-looking nevi are, nonetheless, very common. Moreover, by the time a child reaches adolescence, the incidence of melanoma increases substantially, with a rate of 13.2 cases per million children aged 15-19 years

Discussion

The etiology of congenital melanocytic nevi remains unclear. The melanocytes of the skin originate in the neuroectoderm¹, although the specific cell type from which they derive remains unknown. External insult results in a mutation that affects the morphogenesis of the embryonic neuroectoderm and migration of precursor cells to the skin. Congenital nevi have been stratified into 3 groups according to size. Small nevi are less than 1.5 cm in greatest diameter, medium nevi are 1.5-19.9 cm in greatest diameter, and large or giant nevi are greater than 20 cm in greatest diameter. Giant nevi are often surrounded by several smaller satellite nevi. Neurocutaneous melanosis may result from an error in the morphogenesis of the neuroectoderm², which gives rise to the melanotic cells of both the skin and meninges. Clinically, patients may present with increased intracranial pressure due to hydrocephalus or a mass lesion.¹ The prognosis of patients with symptomatic neurocutaneous melanosis is very poor, even in the absence of malignancy. In one review of 39 reported cases of symptomatic neurocutaneous melanosis, death occurred in more than half the patients within 3 years of the onset of neurological symptoms, and most deaths were in patients younger than 10 years.³

The etiology of congenital melanocytic nevi remains unclear. The melanocytes of the skin originate in the neuroectoderm, although the specific cell type from which they derive remains unknown.²One hypothesis is that pluripotential nerve sheath precursor cells migrate from the neural crest to the skin along paraspinal ganglia and peripheral nerve sheaths and differentiate into melanocytes upon reaching the skin.⁴No racial predilection is recognized for congenital nevi. Congenital nevi occur in both sexes, with no known predilection. To be considered congenital nevi, lesions must be present at birth. Scalp nevi in children younger than 18 years old tend to have perifollicular hypopigmentation that creates the appearance of scalloped, irregular borders if occurring on the periphery, or variegation in pigmentation, if occurring within the nevi

Differential Diagnosis

- Epidermal Nevus Syndrome, Halo Nevus, Malignant Melanoma, Neurofibromatosis, Nevus of Ota and Ito, Nevus, Melanocytic, Nevus Sebaceous, Paget Disease, Mammary, Seborrhic Keratosis, Spitz Nevus.

Conclusion

To be considered congenital nevi, lesions must be present at birth. Scalp nevi in children younger than 18 years old tend to have perifollicular hypopigmentation that creates the appearance of scalloped, irregular borders if occurring on the periphery, or variegation in pigmentation, if occurring within the nevi.

REFERENCE

1. Lemmens OJ, Kroon S. The histology of "congenital features" in early acquired melanocytic nevi. *J Am Acad Dermatol.* Oct 1988;19(4):742-6. | |
2. Krengel S, Hauschild A, Schafer T. Melanoma risk in congenital melanocytic naevi: a systematic review. *Br J Dermatol.* Jul 2006;155(1):1-8. | |
3. An-sarin H, Soltani-Arabshahi R, Mehregan D, Shayanfar N, Soltanzadeh P. Giant congenital melanocytic nevus with neurofibroma-like changes and spina bifida occulta. *Int J Dermatol.* Nov 2006;45(11):1347-50. |
4. Kinsler VA, Abu-Amero S, Budd P, Jackson IJ, Ring SM, Northstone K, et al. Germline Melanocortin-1-Receptor Genotype Is Associated with Severity of Cutaneous Phenotype in Congenital Melanocytic Nevi: A Role for MC1R in Human Fetal Development. *J Invest Dermatol.* May 10; 2012. |