

Pediatrician's Role in the Management of Giant Congenital Melanocytic Nevus: Case Report

Dev Konjenital Melanositik Nevus Yönetiminde Pediatri Hekiminin Rolü

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ABSTRACT Although, congenital giant melanocytic nevi (CGMN) are uncommon in pediatric patients, they carry an increased risk of malignancy, neurocutaneous melanocytosis, and malformations. The management requires a multidisciplinary approach and the pediatrician plays an important role. We aimed to report a case with CGMN, and discuss the management with special emphasis on the pediatrician's role. During a well child visit, physical examination of a 3 months old girl revealed a circumferential large melanocytic nevus over her right leg. There were satellite lesions on her left leg, and back. Melanocytic nevus presents a challenge to the pediatricians because they are lacking the necessary knowledge. Close follow up with documentation of the size, and location of the nevi, palpation of the lesion for nodule, measurement of head circumference and neurodevelopmental assessment, taking an appropriate medical history with emphasis on neurological symptoms associated with neurocutaneous melanocytosis is important.

Key Words: Nevus, pigmented; neurocutaneous melanosis; melanoma

ÖZET Pediatri hastalarında Konjenital Dev Melanositik Nevus (KDMN) nadir görülmekle birlikte, artmış malignite, nörokütanöz melanositoz ve malformasyon riski taşırlar. Yönetimi multidisipliner bir yaklaşım gerektirir ve pediatrist önemli bir rol oynar. KDMN olan bir olgunun sunulması ve pediatristin rolü üzerinde özellikle durularak tedavi yönetiminin tartışılması amaçlandı. Üç aylık kız çocuğun sağlam çocuk randevusunda, fizik muayenede sağ alt ekstremiteyi tamamen çevreleyen geniş melanositik nevus saptandı. Sol bacak ve sırtında da satelit lezyonlar vardı. Melanositik nevus pediatristler için gerekli bilgiye sahip olmadıklarında zorluk yaratır. Yakın izlemle lezyon boyut ve yerinin not edilmesi, nodül açısından palpasyonu, baş çevresi ölçümü ve nörogelişimsel değerlendirme, nörokütanöz melanositoz bulguları açısından uygun tıbbi öykünün alınması önemlidir.

Anahtar Kelimeler: Nevüs, pigmente; nörokütanöz melanozis; melanom

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Congenital melanocytic nevi (CMN) are classified according to the greatest diameter they are expected to attain by adulthood, "projected adult size" (PAS).^{1,2} Krengel et al proposed a new categorization system including size (small <1.5 cm, medium 1.5-20 cm, large 20-40 cm, and giant >40 cm), number of satellite nevi, anatomic localization, and other morphologic features (color, thickness, hairiness).¹ On the other hand, many authors define Giant Congenital Melanocytic Nevi (GCMN) as CMN which measures ≥ 20 cm.^{1,2} Multiple, small CMN located on the head, trunk, and extremities in association with a large CMN are

called satellite nevi.³ Neurocutaneous melanocytosis (NCM) is melanocytic proliferations within the leptomeninges and brain parenchyme in patients having a CMN.^{3,4}

GCMN are rare with an estimated incidence of 1 in 20 000 to 500 000 live births but because of increased risk for developing malignant melanoma, NCM, and rarely other tumors (rhabdomyosarcoma, neurofibroma) and associated malformations (Dandy-Walker malformation, spina bifida occulta) it is an important issue in pediatrics.⁴⁻⁸ Most cases of GCMN are not linked to neurofibromatosis, but up to 5% of patients with neurofibromatosis have been noted to have GCMN.⁷

We aimed to report a case with CGMN and discuss the management of CGMN with special emphasis on the pediatricians' role in the diagnosis, management and follow up.

CASE REPORT

A 3 month old girl was referred to our well child outpatient clinic by the dermatology department for a complete physical examination and follow up. She was born full term from an unrelated family and her postnatal medical history was unremarkable. Her family history was negative for skin disorders. She was born with a large congenital melanocytic nevus and multiple satellite nevi. Her physical examination revealed a circumferential large melanocytic nevus over her right thigh and lower leg measuring 18 cm vertically (PAS>40 cm) in association with underdevelopment of the limb (soft tissue hypoplasia) confined to the region of the nevus, leading to asymmetry and have darkly pigmented hairs on it (Figure 1). The surface of the lesion was rough forming a plaque and the color ranged from dark brown to black. There were satellite lesions on her left leg, middle and lower aspects of back (Figure 2) and medium CMN on her face. No nodules were palpated. The remaining general exam and neurological exam was normal. Leg X-ray showed mild asymmetry with no skeletal abnormalities. Magnetic Resonance Imaging (MRI) of the child's brain and the spine were normal. There were no symptoms of neurocutaneous



FIGURE 1: Circumferential large melanocytic nevus over her right thigh and lower leg with darkly pigmented hairs.



FIGURE 2: Satellite lesions at the lower back of the patient.

melanocytosis. She had no symptoms or malignant transformation during the 12 month follow up period.

DISCUSSION

Our patient had a GCMN with satellite lesions, carrying all the risks associated with increased rate of complications. Patients with larger lesions, lesions located on the back, presence of satellite lesions

have been suggested to be at higher risk for complications.^{5,8-10} GCMN's risk of developing melanoma over lifetime has been reported to be between 2% and 10% with the greatest risk in the first decade.⁵ Kinsler et al.'s prospective study showed that larger lesions and multiple CMNs have increased risk of malignant melanoma especially over the first 2 years.⁸

Neuroradiological screening for NCM is controversial. Some authors don't recommend routine magnetic resonance imaging (MRI) for asymptomatic patients, because they suggest that imaging does not predict which patients will become symptomatic or who will benefit from therapy.³ Also if the patient is symptomatic there is no treatment other than palliative procedures, and the prognosis is poor.^{3,5} On the other hand, Kinsler et al. suggest that the presence of satellite lesions at birth should be the first step in classification.⁸ If satellite lesions were present at birth they recommend gadolinium enhanced MRI of whole central nervous system (CNS) in the first 6 months of life due to myelination after this period.⁸ Hale et al. also demonstrated that there is a significant trend between increased numbers of satellite lesions and the occurrence of both melanoma and NCM.⁴ Since our patient had satellite lesions, we screened whole CNS with MRI and the scans revealed no abnormality.

NCM symptoms often develop within the first few years of life and the patients should be followed for neurologic symptoms like seizures, hydrocephalus, headache, vomiting, increasing head circumference, bulging fontanelle, gait abnormalities, bladder and or bowel dysfunction, cranial nerve dysfunctions and neurodevelopmental delay.^{3,8} In Kinsler's study 20% of the patients had abnormal neurodevelopment and they suggested that all children with CMNs have the potential for abnormal neurodevelopment and neurodevelopmental assessment should be an essential part of the follow up.⁸

Alikhan et al. proposed an algorithm which involves screening for spinal dysraphism for midline lumbosacral CMNs.² It is important to diagnose an occult spina bifida as early as possible for the pa-

tient to benefit from the treatment. Spinal MRI of our patient did not reveal any abnormality.

Lesions are either observed or surgically removed.^{3,9,10} There is no consensus on the management of GCMN. It depends on the location and size of the lesion, parental concerns and the presence of other risk factors. Some authors suggest removal of the lesions as early as possible, due to the increased risk of malignancy in the first years of life and also the elasticity of tissue at young age.^{9,10} On the other hand prophylactic removal is controversial. Kinsler et al suggested that the risk of malignant melanoma is not affected by prophylactic surgery.⁸ It is suggested that malignancy still may occur despite complete excision and also surgical excision will not reduce the risk of extracutaneous melanoma or NCM.^{3,8,10} Also, due to the large size and deep spread of the lesion, complete excision may not be possible.^{3,8,10} Risks of extensive surgery (sepsis, restriction of mobility of the joints due to scarring, poor cosmetic outcomes) and general anesthesia risks should be also considered in patients less than one year of age.^{3,8,10} We referred the patient to plastic surgery; they recommended surveillance only, due to the large size of the lesion where a complete excision of the lesion was not possible. They invited the patient at the end of the first year for reevaluation.

Many authors recommend three monthly visits for those with satellites and six monthly visits with large CMN without satellites in the first 2-4 years of life, the time when the risk of developing melanoma is highest, and yearly thereafter.^{3,8} We performed monthly visits at first and then recommended three monthly visits thereafter. We gave special emphasis on neurological symptoms, head circumference measurements, neurodevelopmental status of the patient. Melanoma developing within GCMN may develop deeper within the tissue which makes it harder to detect with visual inspection only.^{3,4,8} Parental self examination is also important for the same reason. The parents were also taught about how to palpate the lesion and warned about neurological symptoms. The parents also should be educated about observing the progressive changes in appearance of the nevi, like

asymmetry, irregular border, colour change, amelanosis, increasing lumpiness, bumps, bleeding and development of new satellite changes.^{11,12} Recent evolution of the lesion has been suggested as universally valuable.¹² If concerning features is observed referral to an experienced dermatologist is required. However it should also be noted that the concerning features is not as helpful in children as in adults and prognostic factors are less clear in

children.¹² In our patient, the parents were more anxious about the patient's cosmetic appearance rather than increased risk of malignancy. Psychological support is also provided to the parents.

In conclusion, wait and see approach is becoming more popular each day and the pediatrician must know her role in the multidisciplinary approach which involves the pediatrician, dermatologist, plastic surgeon, and the psychologist.¹⁰

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