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Multiple Giant Congenital Nevi – A case report

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VERSITA

Abstract

Giant congenital melanocytic nevi are benign nevomelanocytic proliferations of 20 cm or more in diameter, present at birth. They are primarily found on the posterior trunk, but they may arise on any other part of the body, covering more than 2% of the body surface. Giant congenital nevi are major risk factors for the development of melanoma, and the risk has been estimated to be as high as 5–7%. Persons with giant congenital melanocytic nevi on the head, neck and along the midline of the back are at increased risk for leptomeningeal melanocytic lesions. Most patients with neurocutaneous melanosis present with neurologic manifestations of the disease in the first 2 years of life. Melanoma occurs in 62–80% of cases, but even without neoplasms, symptomatic neurocutaneous melanosis has a poor prognosis. This is a report of a 23-year-old female patient who presented with multiple congenital pigmented and pilous nevi covering over 2% of her total body surface, without malignant alterations or association with other abnormalities. At birth, a nevus covered her neck, shoulders and the upper left arm, whereas several nevi over 5cm in diameter were present in the gluteal region, on the abdomen and legs. During the first 2 years of life, the existing nevi increased in size and progressed into darker brown. New, smaller pigmented changes appeared on the whole body and the face, while at the age of 17 they reached their current size and layout. At puberty, nevi over 10cm in size grew dark hairs. There were neither melanoma nor skin tumor cases in the family. Nuclear magnetic resonance imaging was not performed in the childhood or later in life, but other parameters – neurologic and ophthalmologic findings were in normal range all the time, as was growth and development. A complete photo-documentation was made, including macroscopic and dermoscopic images and regular follow-ups continue.
Giant congenital melanocytic nevi may cause considerable esthetic and psychosocial problems. Due to their high malignant potential, association with other abnormalities, no consensus on the treatment, and monitoring problems, giant congenital melanocytic nevi represent a therapeutic problem as well.

Key words

Nevus, Pigmented + congenital; Skin Neoplasms + congenital; Neurocutaneous Syndromes + congenital; Melanoma + etiology; Skin Abnormalities; Abnormalities, Multiple

Giant congenital melanocytic nevi (GCMN) are benign nevomelanocytic proliferations of 20 cm or more in diameter, present at birth (1). They are primarily found on the posterior trunk, but they may arise on any other part of the body, covering over 2% of the total body surface (2). The appearance of GCMN may change over time: the color may get darker, with dark thick pigmented hairs, whereas the surface may be smooth, wrinkled, verrucous or cerebriform. Satellite nevi of different sizes surround the GCMN. Several developmental abnormalities have been reported to be associated with GCMN: scoliosis, spina bifida, clubfoot, elephantiasis, cranial bone hypertrophy, and neorocutaneous melanosis (3).

Congenital nevi are major risk factors for melanoma (2, 3, 4). Fortunately, melanoma remains an uncommon malignancy in children aged 0-9 years, with an annual incidence of 0.7 cases per 1 million children. For GCMN, the risk has been estimated to be as high as 5–7%. However, in cases with GCMN during the first 15 years of life, this risk increases up to 8.52%. GCMN may cause considerable esthetic and psychic problems.

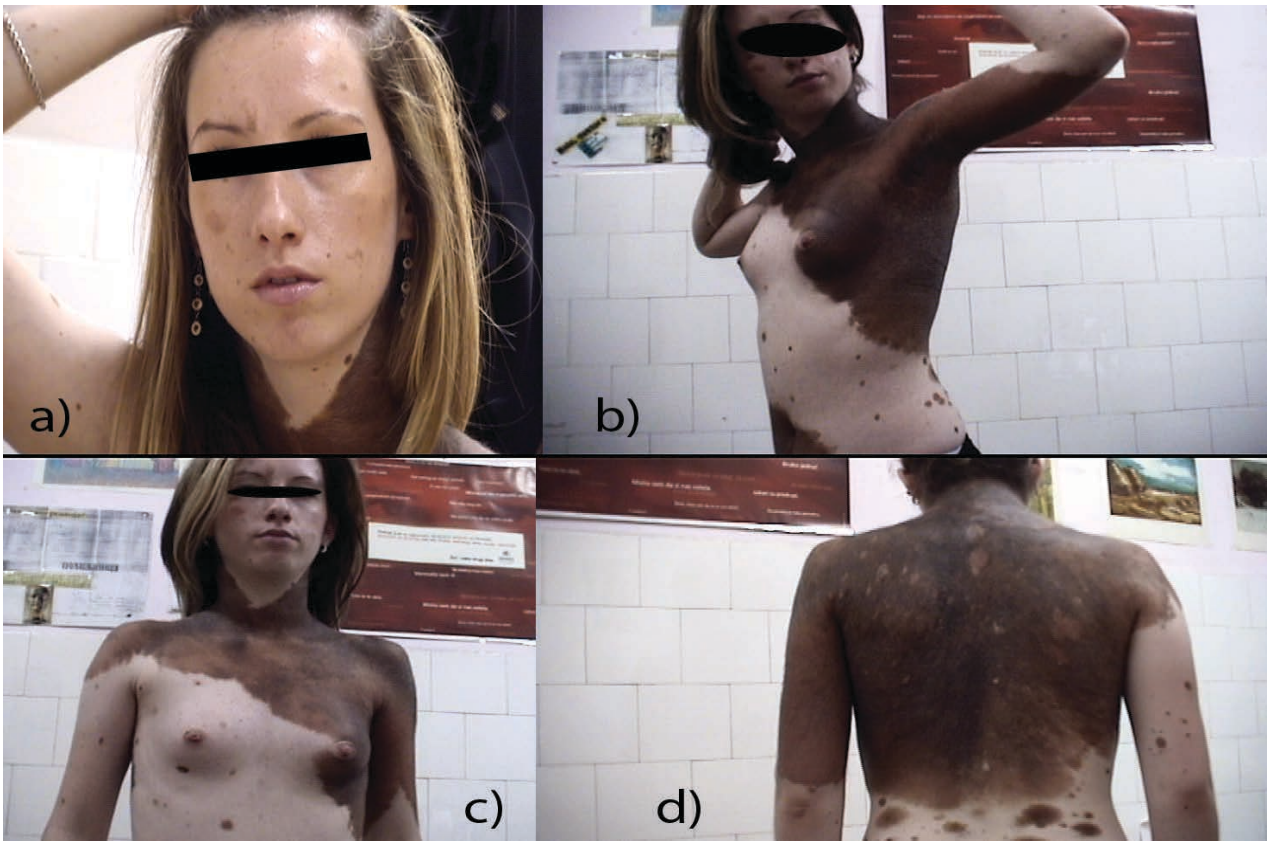

Figure 1. A dark brown nevus covering a) the entire neck; b) both shoulders, left upper arm, left breast; c) anterior thorax from the right shoulder to the left rib cage; d) the entire back up to the waist

This is a report of a female patient with nevi affecting over 2% of the total body surface, which makes a problem in controlling the patient properly.

Case report

This is a report of a 23-year-old female patient who presented with multiple congenital pigmented and pilous nevi, covering over 2% of her total body surface. At birth, a giant nevus covered her neck, shoulders and the upper left arm, whereas several nevi over 5cm in diameter were present in the gluteal region, on the abdomen and legs. During the first 2 years of life, the existing nevi increased in size and progressed into darker brown. New, smaller pigmented changes appeared on the whole body and the face, while at the age of 17 they reached their current size and layout. At puberty, nevi over 10cm in size grew dark hairs. There were neither melanoma nor skin tumor cases in the family.

During the first years of life, the patient was controlled by a pediatrician, a neurologist and an ophthalmologist. Although the patient was diagnosed with a giant congenital nevi, magnetic resonance imaging (MRI) of the head was not performed in the first two years of life. Fortunately, she had no signs and symptoms of increased intracranial pressure and spinal cord compression. Growth and development was in normal range, so annual monitoring by a neurologist and an ophthalmologist was continued.

On the first examination in 2007, more nevi were observed: a dark brown nevus of unclear edges, partially with thick dark hairs, covered the entire neck circumference (except for a v-shaped part on the lower jaw), both shoulders, the left upper arm, left breast, anterior thorax – from the right shoulder to the left rib cage, and the entire back up to the waist (Figures 1a, 1b, 1c, 1d).

The skin of the gluteus and extremities (Figures 2a, 2b, 2c 2d), as well as of the right sole and the left foot (Figures 3a, 3b), was covered by a total of over 100 nevi, mostly oval in shape, of uniform light

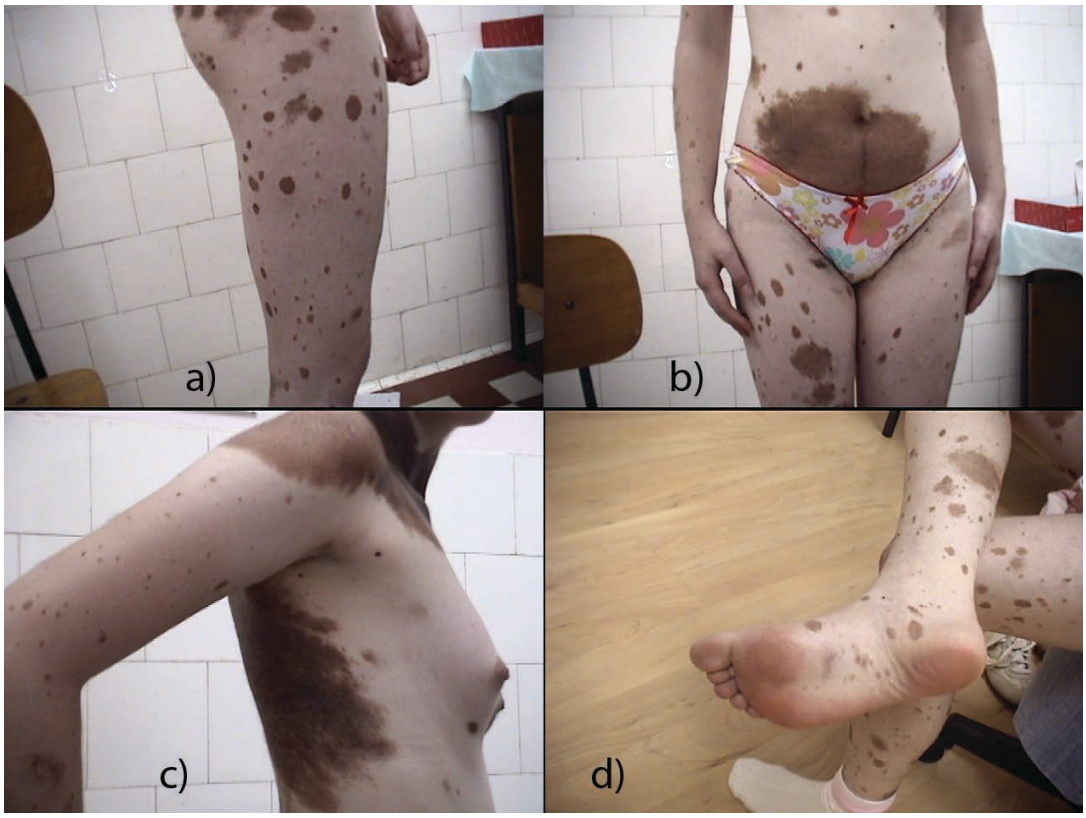

Figure 2. A total of over 100 nevi, mostly oval in shape, of uniform light to dark brown color from 0,5 to 26cm in size covering the skin of the: a) right buttock; b) lower abdomen and both upper legs; c) right arm; d) righ sole and lower leg

to dark brown color, some growing hair, from 0.5 to 26cm in size (Figures 4a, 4b)**.**

Dermoscopy revealed a homogenous brown pigmented network with oval shaped skin-colored macules, with regular distribution corresponding to follicular ostia. The pigmentation of the edge of ostia was marked and symmetrical, indicating benign pigment changes (Figure 5). Apart from a homogenous, a globular pigment network was found in some parts of the nevus, resembling paving stone,

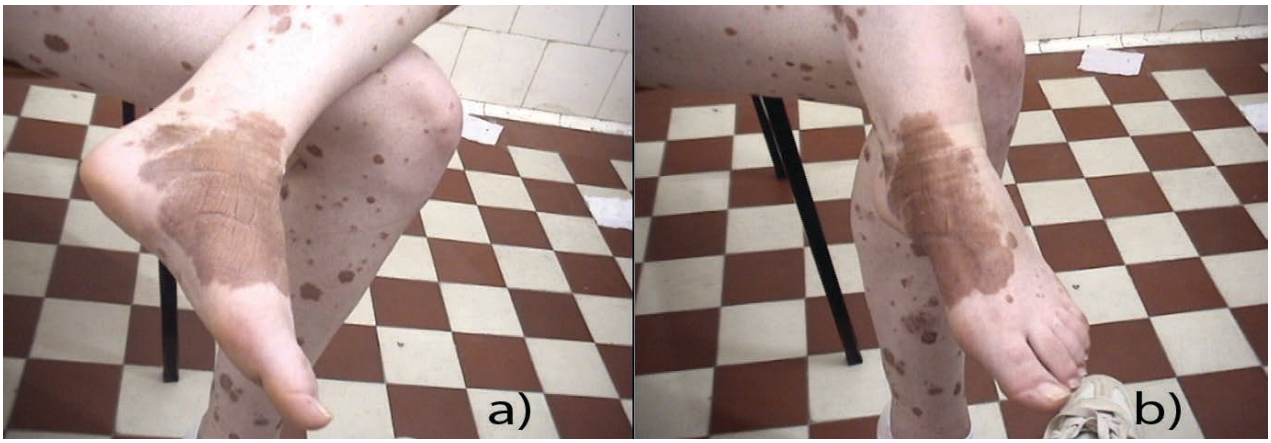

Figure 3. A light brown nevus covering the skin of the left foot: a) from the entire dorsum; b) to the inner sole.

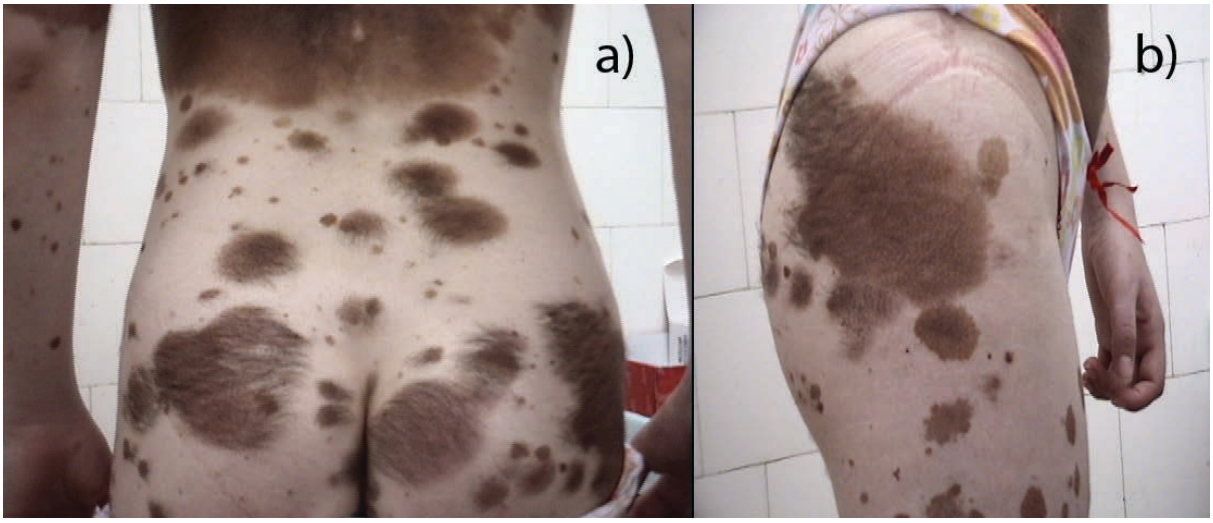

Figure 4. Dark brown nevi growing hair covering the skin on the: a) gluteal region; b) right buttock

which is a typical finding in congenital nevi (Figure 6). Individual, small to medium sized nevi presented with a globular brown network. Some of them had globules of the same size and shape, distributed on the edges, pointing to the growth of changes. Palmar and plantar nevi were monochrome, with parallel and lattice-like patterns, and of benign type (Figures 7a, 7b).

A photo-documentation was made, including macroscopic whole-body images, and dermoscopic images of some nevi and parts of nevi.

Discussion

In the evaluation of patients with GCMN, it is necessary to exclude association with other abnormalities and syndromes, as well as differential diagnosis of other pigment changes, such as:

Neurocutaneous melanosis (NCM) is a rare congenital syndrome characterized by the presence of large and multiple congenital melanocytic cutaneous nevi and benign or malignant melanotic neoplasms. Persons with large GCMN on the head, neck and along the midline of the back are at increased risk of leptomeningeal melanosis. The syndrome is probably an error in the morphogenesis of embryonal neuroectoderm. Most patients with neurocutaneous melanosis show neurologic manifestations of the disease in the first two years of life. Melanomas occur in 62–80% of cases, but even without neoplasms, symptomatic neurocutaneous

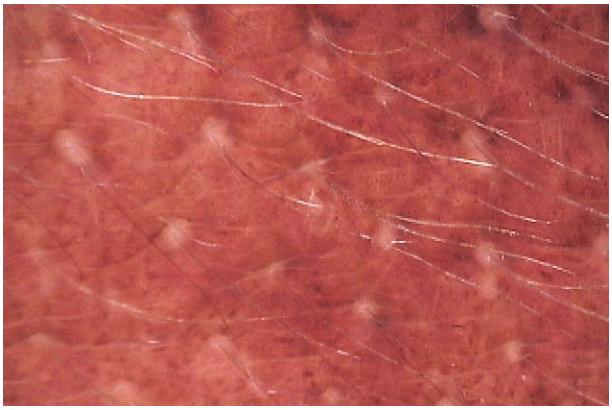

Figure 5. Dermoscopy of the dark brown nevus on the back revealing a homogenous brown pigmented network with oval shaped skin-colored macules with regular distribution corresponding to follicular ostia; the pigmentation of the edge of ostia is marked and symmetrical

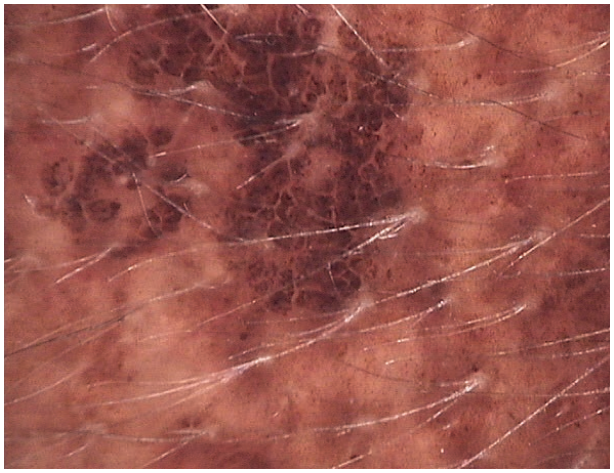

Figure 6. Dermoscopy of the dark brown nevus on the back revealing a globular pigment network in some parts of the nevus, typically resembling paving stone

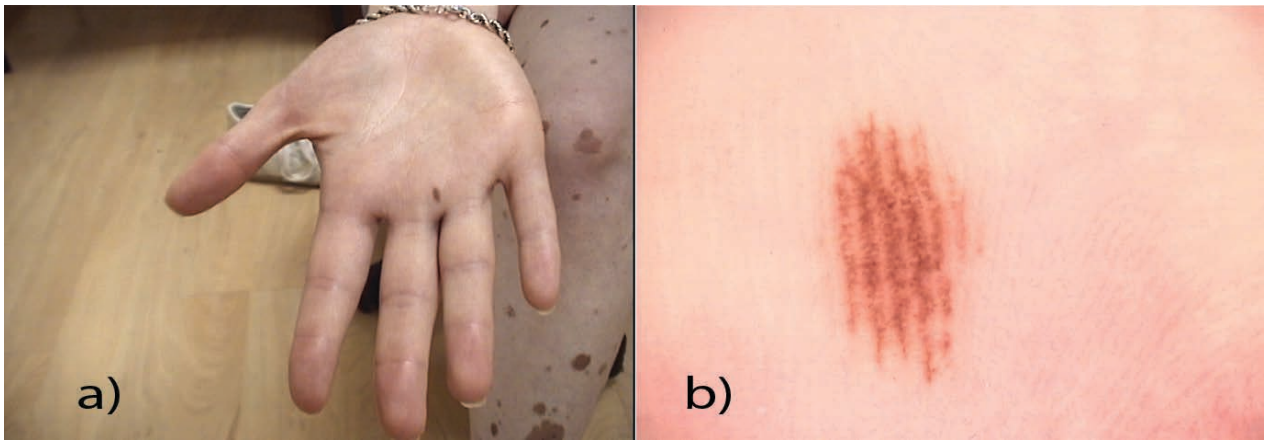

Figure 7. a) Light brown nevus on the left palm; b) dermoscopy showing parallel and "lattice-like patterns"

melanosis has a poor prognosis. MRI of the head and neck is recommended until the second year of life, in order to establish leptomeningeal melanosis, whereas after that a follow up of neurological signs and symptoms pointing to increased intracranial pressure should be done. (4,5,6).

Nevus of Ito is a dermal melanocytic nevus affecting the shoulder area. It is often associated with nevus of Ota, which commonly involves the ocular area – innervated by the trigeminal and maxillary nerves. Both nevi are rare in Caucasians, but common in Asian populations, 0.2-0.6%. Nevus of Ito is an esthetic problem. To date, only one case of melanoma has been described as nevus of Ito, in a 78-year-old male Caucasian (7).

Becker's nevus is a pigmented hairy epidermal nevus. It is more common in male population. It initially presents with an asymptomatic macula, irregular in shape, tan-to-brown in color, most commonly located on the shoulder, upper back and the thorax, in the first 15 years of life, but not at birth. With time, the pigmentation keeps spreading, and hairs start growing (8).

Giant congenital melanocytic nevi may lead to esthetic and psychological problems. Due to their high malignant potential, association with other abnormalities, no consensus on the treatment, and monitoring problems, GCMN is a therapeutic problem as well

Whether treatment should be initiated (2, 3), and what type of treatment it should be depends on many factors: size of the nevus, localization, cosmetic effects, age, and risk of general anesthesia (9). Although the risk of malignant alterations of small and medium congenital nevi has not been established, most dermatologists believe that the risk is not so great as to perform a prophylactic removal of these nevi. Literature data show that there are more and more physicians speaking in favor of prophylactic excision of nevi until puberty, because it is the time when the risk of developing melanoma is greatest. Assessing the risk of melanoma, of general anesthesia, and psychological factors, the best time for excision is between the 6th and the 9th months, and between the 8th and 12th years of life.

Therapeutic modalities include all means which may reduce the melanocyte count that theoretically reduces the risk of cutaneous melanoma: removal of the full depth of nevus, partial excision of nevus, curettage, dermabrasion, laser therapy, and chemical peeling (4). However, apart from complete excision of nevus, other therapeutic modalities do not affect the risk of melanoma in the deep layers of the nevus. For example, superficial nevus destruction by laser results with minimal scarring, and that is why it is most acceptable for patients, but due to insufficient tissue removal, it causes nevus recurrence (4,10). Effects of sublethal laser energy treatment on melanocytes, in regard to malignant alterations, are unfamiliar, but after laser therapy, no malignant alterations have been reported. Furthermore, if melanoma appears on the site of nevus treated by laser, it will be deep in the tissue, which limits the clinical diagnosis, until it reaches an advanced stage. Nevertheless, even total excision of GCMN will not entirely remove the risk of extracutaneous melanoma.

In places that are visible and excision is difficult, application of corrective cosmetics is possible, to

achieve a satisfactory esthetic appearance. Otherwise, a combination of dermabrasion/peeling and corrective cosmetics is recommended.

If focal growth, with changes in color or texture, as well as local sensitivity or ulcerations, potential signs of melanoma, is noticed, biopsy and histopathology analysis are necessary. Also, recommendations for rigorous sun protection and self-examination once a month are crucial.

Proper follow-up of patients with GCMN includes the following procedures:

1. Newborn infants with giant congenital nevi (4) should undergo MRI of the head, in order to detect NCM in the first 4 months of life, before myelinization is complete, because it has been observed that myelin may obscure deposits of melanocytes in the leptomeninges. MRI should be repeated in the first 2 years of life, regardless of the presence or absence of neurological symptoms, because the peak incidence of extracutaneous melanomas and of other tumors is by the second year of life. The necessity of MRI later in life has not been proven, especially in people with regular neurological status and normal development (4).
2. In patients with GCMN and regular neurological status, without atypical findings, periodical follow-up of nevi is recommended, with photo-documentation including macroscopic and dermoscopic images, whereas neurological examinations and sun protection are necessary.
3. In patients with GCMN, regular neurological status and presence of atypical findings, biopsy and histopathological analysis are recommended. If alterations correspond to melanoma, total excision should be considered, or excision with appropriate margins for tumor thickness, as well as regular follow-ups according to the protocols for monitoring melanoma patients.
4. In patients with GCMN and regular neurological status, but with MRI findings pointing to NCM (asymptomatic NCM), frequent MRI is recommended, as well as further therapeutic measures, depending on the disease progression.
5. In patients with symptomatic NCM, neurosurgery may be elected: ventriculo-peritoneal shunt placement may significantly improve the condition in some patients, but also enable migration of melanocytes from leptomeninges to the peritoneal cavity. Patients with symptomatic NCM, whose symptoms have improved, should undergo regular MRI and neurological examinations.

Despite the fact that most patients with GCMN will never develop melanoma, the relative risk is rather high and it should be taken into consideration when evaluating these patients. The decision on the choice of treatment should be left to the patient, after good assessment of risks and benefits of each therapeutic procedure.

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Abbreviations

GCMN - Giant congenital melanocytic nevi
NCM - Neurocutaneous melanosis
MRI - Magnetic resonance imaging

Multipli gigantski kongenitalni nevusi – prikaz slučaja

Sažetak

Uvod: Gigantski kongenitalni melanocitni nevusi (GKMN) su benigne nevomelanocitne proliferacije veličine od 20 cm i više u dijametru, prisutne na rođenju.1 Najčešće su lokalizovani na zadnjem delu trupa ali mogu biti prisutni na bilo kom delu tela zahvatajući od 2% od ukupne površine tela i više. GKMN se smatraju jednim od faktora rizika za nastanak melanoma (M), i to 8,52% u toku prvih 15 godina života. Procenjen životni rizik za razvoj melanoma kod GKMN iznosi 5-7%.

Nekoliko razvojnih anomalija je udruženo sa GKMN: skolioza, spina bifida, kriva stopala, elefantijaza, hipertrofija kranijalnih kostiju i neurokutana melanoza3 (NKM).Neurokutana melanoza je redak kongenitalni sindrom koga karakteriše prisustvo velikih ili multiplih kongenitalnih melanocitnih nevusa i benignih ili malignih melanotičnih neoplazmi leptomeninga. Osobe sa velikim GKMN na glavi, vratu i duž srednje leđne linije imaju povišen rizik zahvaćenosti leptomeninga. Većina pacijenata sa neurokutanom melanozom već u prve 2 godine života dobije neurološke pritiska. manifestacije bolesti. Melanom se javlja u 62% - 80% slučajeva, ali i bez prisutne neoplazme simptomatska neurokutana melanoza ima lošu životnu prognozu. Preporuka je da se uradi MR glave i vrata do 2. godne života radi dokazivanja postojanja leptomeningealne melanoze, a kasnije nastavi sa praćenjem pojave neuroloških simptoma i znakova povišenog intrakranijalnog pritiska.

Prikaz slučaja: Prikazujemo osobu ženskog pola, 23 godine, sa mnogobrojnim kongenitalnim nevusima koji zahhvataju više od 2% površine tela kod koje nije primeća maligna alteracija i udruženost sa drugim anomalijama. Magnetna rezonanca nije radjena u detinjstvu, ali su ostali parametri – neurološki i oftalmološki nalazi bili u granicama normale, kao i rast i razvoj. Napravljena je kompetna foto dokumentacija uključujući makro snimke nevusa i dermoskopske snimke, i kontrole se obavljaju perodično.

Diskusija: Gigantski kongenitalni melanocitni nevusi predstavljaju kozmetski i psihosocijalni problem pacijentima. S obzirom na visok maligni potencijal i udruženost sa drugim abnormalnostima, nepostojanja konsezusa po pitanju terapijskog pristupa, teškoćama pri monitoringu pacijenata, GKMN predstavlja terapijski problem i lekarima.

Da li će se preduzeti terapija2,3, i kakva će ona biti zavisi od mnogo faktora: veličine nevusa, lokalizacije, kozmetskog efekta, godišta pacijenta i rizika od opšte anestezije.9 Mada rizik maligne alteracije kod malih i srednjih kongenitalnih nevusa nije utvrđen, većina dermatologa smatra da rizik nije tako veliki da bi se pristupilo profilaktičkom uklanjanju ovih kongenitalnih nevusa. Za GKMN se sve više u literaturi pojavljuju stavovi koji govore u prilog profilaktičke ekscizije nevusa, i to do puberteta jer je najveći rizik za razvoj melanoma upravo do tada. Procenjujući rizik za razvoj melanoma, rizik od opšte anestezije i psihosocijalne faktore, smatra se da je najbolji period za eksciziju GKMN izmedju 6. i 9. meseca života i izmedju 8. i 12. godine.

Terapijski modaliteti4 uključuju sve načine kojima je moguće smanjiti broj melanocita što teoretski treba da smanji rizik nastanka kutanog melanoma: ekscizija kompletne debljine nevusa, delimična ekscizija nevusa, kiretaža, dermabrazija, laserska terapija i hemijski piling. Medjutim, osim kompletne ekscizije nevusa, ostali načini terapije ne utiču adekvatno na rizik od nastanka melanoma u dubljim slojevima nevusa, ali čak i totalnom ekscizijom GKMN ne anulira se mogućnost nastanka ekstrakutanog M.

Na mestima koja su vidljiva, a ekscizija je teško izvodljiva, moguća je upotreba korektivnih kozmetičkih preparata radi postizanja zadovoljavajućeg estetskog izgleda ili kombinacija dermabrazije/pilinga i korektivne kozmetike.

Tok i kontrole: Uzimanje biopsije uz sledstvenu histopatološku analizu predstavlja postulat ukoliko se primeti fokalni rast sa promenom boje i teksture, pojava lokalne osetljivosti ili nastanka ulceracije, što predstavlja znake mogućeg razvoja melanoma. Takođe, neophodan je savet za rigoroznu zaštitu od sunca i samopregledi 1 x mesečno.

Adekvatna kontrola pacijenata sa GKMN obuhvata sledeće postupke:

1. novorođenčad sa gigantskim kongentialnim nevusima 4 treba uputiti na magnentnu rezinancu (MR) glave radi otkrivanja asimptomatske NCM u toku prva 4 meseca života, pre završetka mijelinizacije jer je primećeno da mijelin može da prikrije postojanje depozita melanocita na leptomeningama. MR treba ponoviti u toku prve dve godine života bez obzira na prisustvo ili odsustvo neuroloških simptoma jer je pik incidence ekstrakutaanog melanoma i drugih tumora upravo do druge godine života. Neophodnost ispitivanja MR u kasnijem životnom dobu nije dokazana, naročito kod osoba sa urednim neurološkim statusom i normalnim razvojem4;
2. Kod pacijenata sa GKMN i urednim neurološkim statusom, bez prisustva ,,atipičnih" polja preporučuje se periodična kontrola nevusa uz foto dokumentaciiju;

3. periodične kontrole koje podrazumevaju makroskopske i dermoskopske slike, neurološki pregled i preporuke o zaštiti od sunca;
4. kod pacijenata sa GKMN i urednim neurološkim statusom, i prisustnim ,,atipičnim" poljem preporučuje se biopsija i histopatološka analiza. Ukoliko promena odgovara melanoma, potrebno potrebno je razmotriti totalnu eksciziju nevusa ili eksciuiju sa odgovarajućim amrginama za debljinu tumora i redovne kontrole prema protokolu praćenja za melanome;
5. Kod pacijenata sa GKMN i urednim neurološkim statusom, a MR sugestivnim na NKM (asimptomatska NKM) preporučuju se učestale kontrole MR i zavisno od napredovanja bolesti dalje terapijske mere;
6. Kod pacijenata sa simptomatskom NKM moguća je neurohiriška terapija kao što je postavljanje ventrikuloperitonealnog šanta koji može značajno da popravi stanje kod nekih pacijenata, ali i omogući migraciju melanocita sa leptomeninga u peritonealnu duplju;
7. pacijente sa simptomatskom NKM čiji se simptomi popravljaju treba dalje kontrolisati uz ponavljane MR i neurološke preglede.
Zaključak: Relativni rizik za razvoj melanoma je veliki i ne treba ga podceniti uprkos činjenici da većina pacijenata sa GKMN neće nikada dobiti melanom, relativan rizik za razvoj melanoma je veliki i treba ga imati u vidu u evaluaciji ovih pacijenata. Odluku o terapijskom izboru treba prepustiti pacijentu posle dobrog upoznavanja sa rizikom/benefitom svake terapijske procedure ponaosob.

Ključne reči

Pigmentni nevus + kongenitalni; Neoplazme kože + kongenitalne; Neurokutani sindromi + kongenitalni; Melanom + etiologija; Abnormalnosti kože; Multiple abnormalnosti