



Nevus de sitios especiales



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Nevus de Sitio Especial

“Grupo dentro del espectro de los nevus presentes en ciertas localizaciones anatómicas, que tienen características preocupantes histológicamente, pero un comportamiento benigno”

Nevus benignos con atipia histológica similar al melanoma o nevus displásico

Nevus de Sitio Especial

Espectro histológico de lesiones melanocíticas en los sitios especiales:

- Nevus melanocitico pequeño (<5mm)
- Nevus lentiginoso
- Nevus displásico
- Melanoma lentiginoso
- Lentigo Maligno
- Nevus de sitio especial

Nevus de Sitio Especial

- Reflejan diversidad regional en estructura y función de la piel
- Etiología controversial, desarrollo embriológico
- Pueden tener características histológicas comunes con melanoma

Localizaciones

- Línea mamaria embrionaria (seno, axila, ombligo, genitales)
- Áreas flexurales
- Superficies acrales
- Oreja
- Cuero cabelludo

Table 1
Nevi of special sites

| Special Site | Locations |
|---------------|---|
| Genital nevi | <ul style="list-style-type: none">• Vulva• Penis^{7,a} |
| Flexural nevi | <ul style="list-style-type: none">• Nevus of the milkline<ul style="list-style-type: none">◦ Axillae◦ Breast◦ Inguinal folds• Other flexural sites<ul style="list-style-type: none">◦ Umbilicus◦ Scrotum◦ Perianum |
| Acral nevi | <ul style="list-style-type: none">• Ears• Palms• Soles• Knees^{15,a}• Elbows^{15,a} |
| Scalp | |

^a Anecdotal reports only.

Nevus de Sitio Especial

Nomenclatura no estandarizada:

- AGN: *nevus genitales atípicos*
- AMNGT: *Nevus melanocíticos atípicos tipo genital*
- MANIACS: *Nevus melanocíticos especiales con ascenso de células intraepiteliales.*
- *Nevus lentiginoso acral de la piel plantar*
- *Nevus atípico del cuero cabelludo*

Nevus Genital

- Frecuentes en la infancia, <20 años.
- 2 % ♀ tienen nevus vulvares.
- 5% son de sitio especial
- Labios, clítoris y periné
- Igual distribución en áreas mucosas o con pelo.
- > Mucosas en menores de 10 años.

Table 2
Clinical appearance of nevi of special sites

| Special Site | | |
|---|---------------------------|--|
| Synonyms | Location | Clinical Appearance of NOSS |
| Genitalia <ul style="list-style-type: none">• Atypical genital nevi (AGN)⁶• Atypical melanocytic nevus of the genital type (AMNGT)⁷ | Unspecified | <ul style="list-style-type: none">• Black (one-third)⁷• Brown (two-thirds)⁷ |
| | Labia majora and perineum | <ul style="list-style-type: none">• Elevated⁷• Mushroom-shaped⁷ |
| | Labia minora and clitoris | <ul style="list-style-type: none">• Macular⁷ |

Nevus Genital



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Histología

Proliferación prominente de nidos ovales o redondos en la unión, pueden ser confluentes.

Tres patrones:

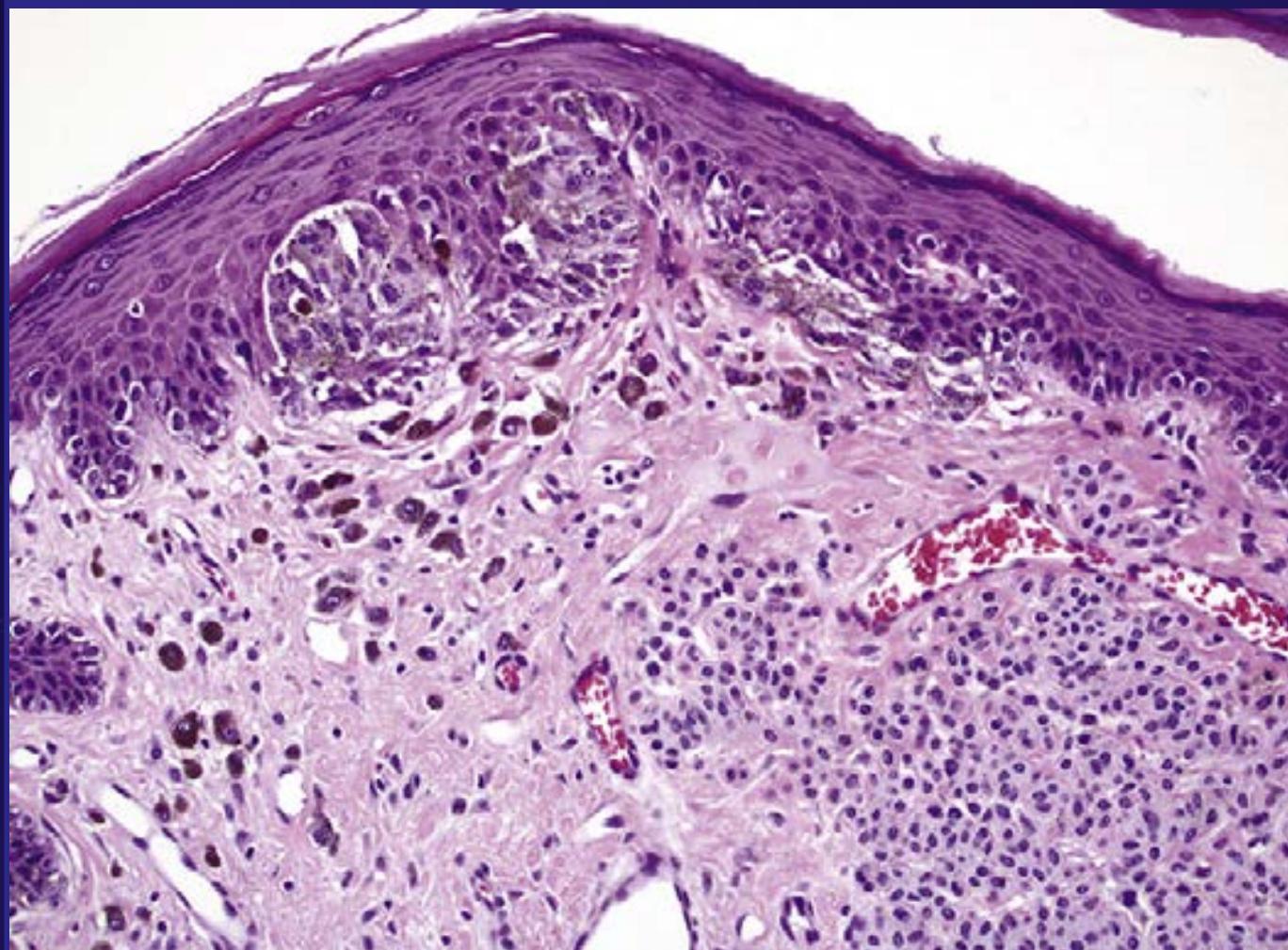
- Patrón anidado: grandes nidos ovales orientados perpendicularmente o paralelos a unión dermoepidérmica.

Histología

- Patrón de nido poco cohesivas: Nidos poco cohesivos forman una banda que separa la epidermis de melanocitos dérmicos maduros
- Patrón Apiñado: Nidos mal definidos y células individuales están estrechamente adosadas, oscureciendo la unión dermo-epidérmica

Histología

Patrón anidado

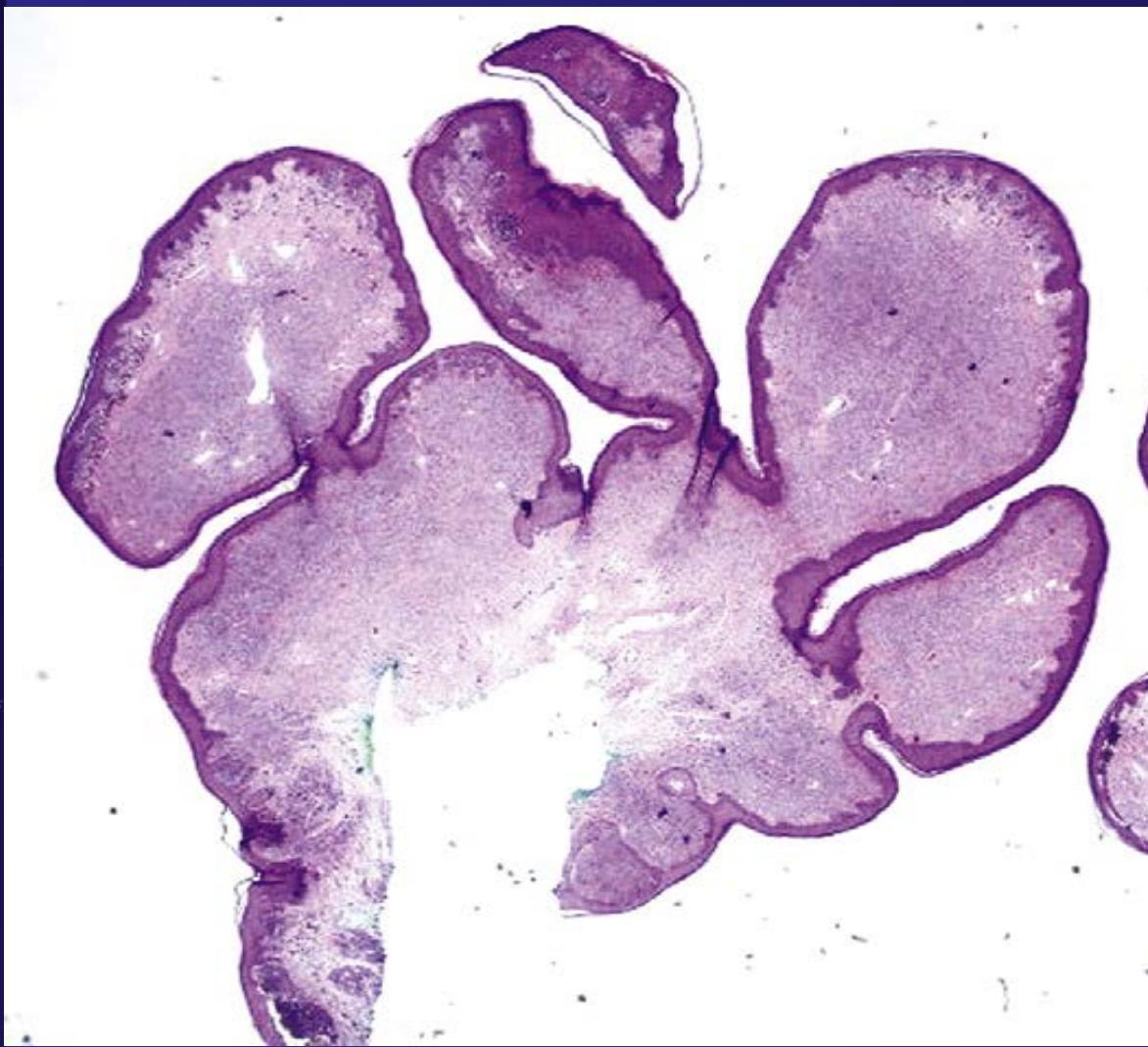


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Histología

- Atipia citológica difusa moderada a severa
- Células epitelioides con nucleolos prominentes
- Diseminación pagetoide focal central
- Amplia zona de fibrosis dérmica

Histología



Prominente morfología
en forma de hongo
evidente a bajo aumento

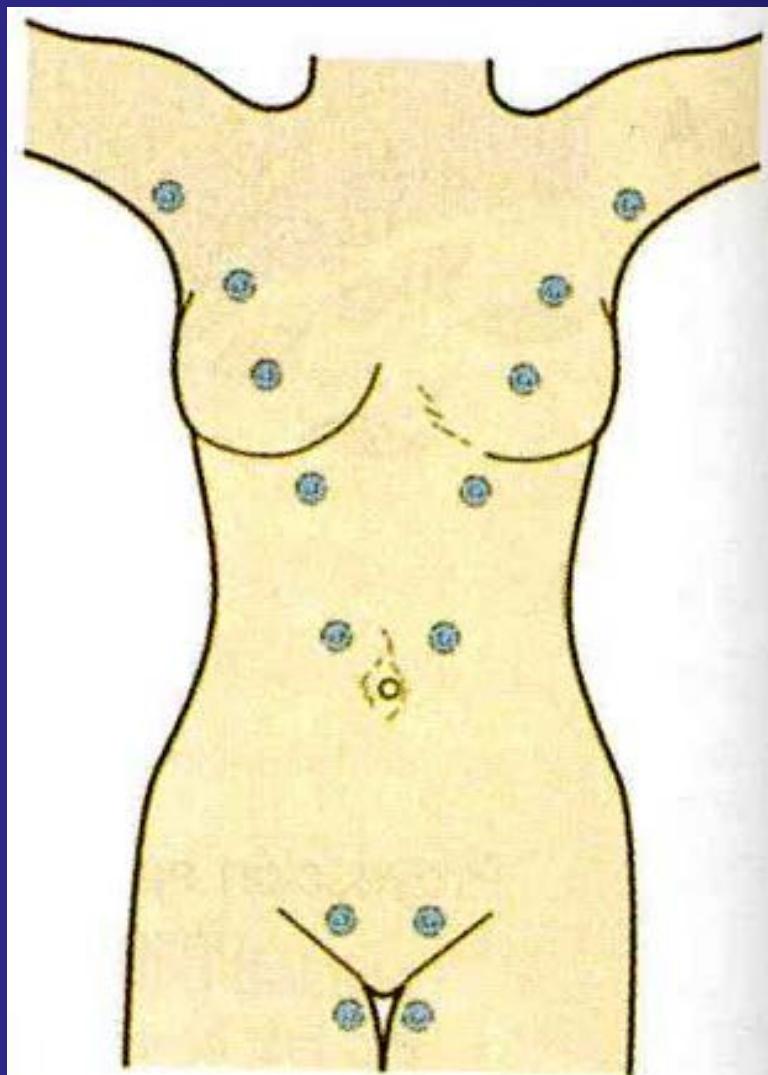
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Histología

Características de benignidad:

- Simetría
- Bordes bien delimitados
- Maduración dérmica
- Baja actividad mitótica dérmica

Nevus de Línea Mamaria Flexurales



- Ombligo (40%)
- Axilas (32%)
- El resto: pliegue inguinal, pubis, escroto y region perianal.

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Table 2
Clinical appearance of nevi of special sites

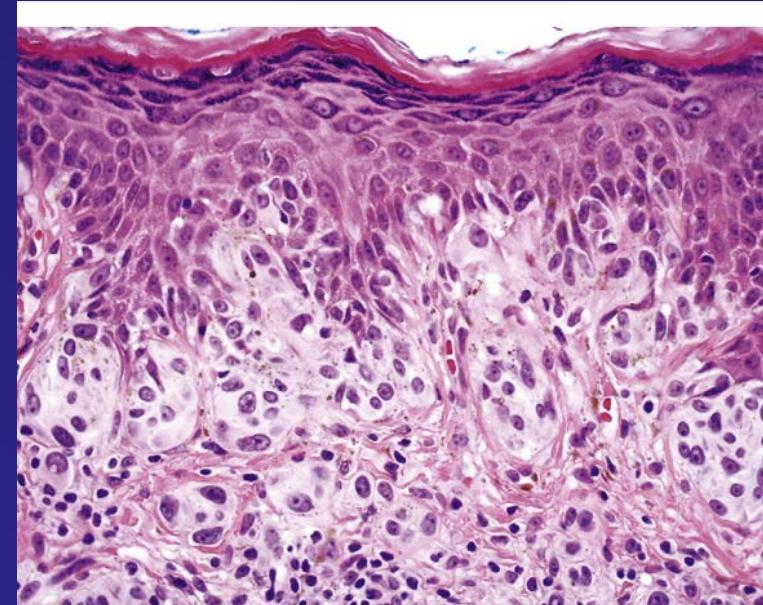
| Special Site | Location | Clinical Appearance of NOSS |
|---|---------------------------|---|
| • Synonyms | | |
| Genitalia | Unspecified | <ul style="list-style-type: none"> • Black (one-third)⁷ • Brown (two-thirds)⁷ |
| <ul style="list-style-type: none"> • Atypical genital nevi (AGN)⁶ • Atypical melanocytic nevus of the genital type (AMNGT)⁷ | Labia majora and perineum | <ul style="list-style-type: none"> • Elevated⁷ • Mushroom-shaped⁷ |
| | Labia minora and clitoris | <ul style="list-style-type: none"> • Macular⁷ |
| Flexures | Milkline | <ul style="list-style-type: none"> • No data |

Nevus de Línea Mamaría Flexurales

- Comparte características histológicas con los nevus genitales:
 - Fibroplasia en la dermis papilar
 - Maduración
 - Proliferación lentiginosa melanocitaria.
 - Extensión del componente intradérmico a lo largo de anexos
 - Patrón anidado con células poco cohesivas

Nevus de Línea Mamaría Flexurales

- Nevus de los senos son histológicamente diferentes:
 - Melanocitos intraepidérmicos por encima de la capa basal, individuales o en grupos.
 - Fibroplasia de la dermis papilar
 - Atípia melanocitaria



Nevus Acrales

- Plantas
- Palmas
- Orejas
- Región retroauricular
- Región preauricular
- Mejilla

Table 2
Clinical appearance of nevi of special sites

| <u>Special Site</u> | | |
|---------------------|--|---|
| <u>Synonyms</u> | <u>Location</u> | <u>Clinical Appearance of NOSS</u> |
| Acral skin | Soles | <ul style="list-style-type: none"> • Brown to dark brown macules¹⁹ • Small (<10 mm)¹⁹ • Uniform in color¹⁹ • Regular borders¹⁹ |
| | Palms | <ul style="list-style-type: none"> • No data |
| | Auricular region, ²¹ external auricle, ²⁰ postauricular cheek, ²⁰ and preauricular skin ²⁰ | <ul style="list-style-type: none"> • Symmetric²¹ • Ill-demarcated²¹ • Approximately half are at least 6 mm²¹ |

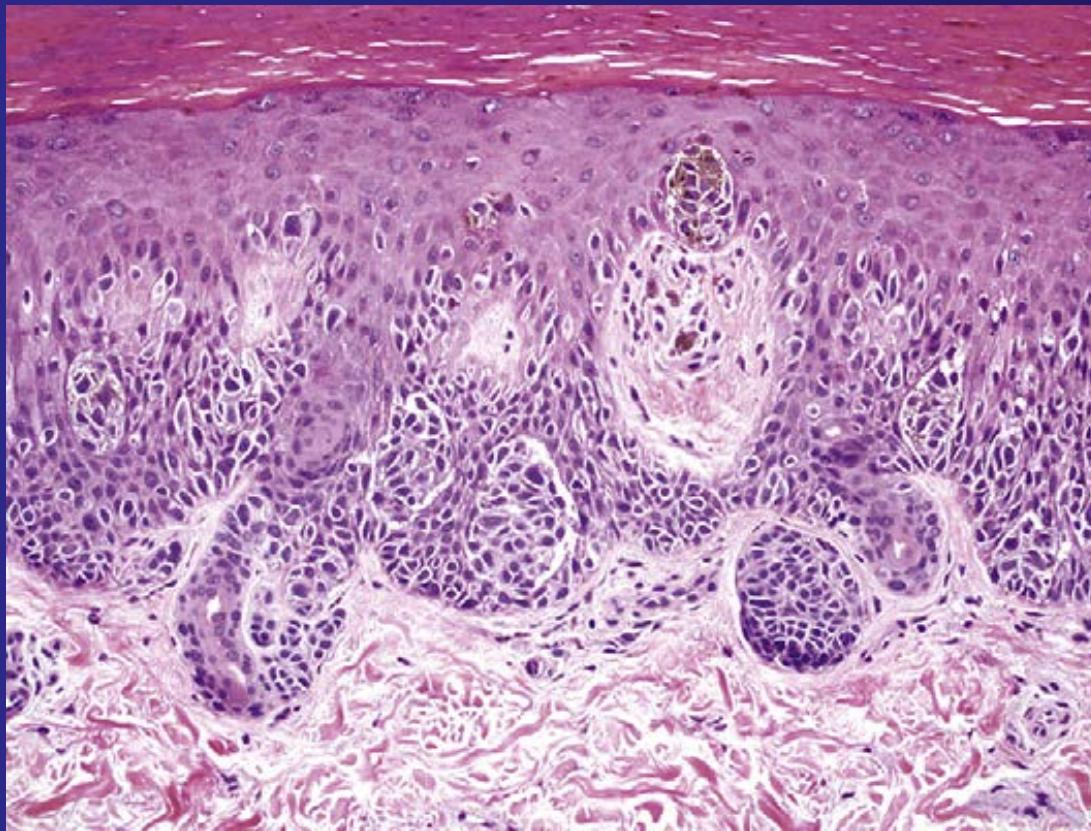
Nevus Acrales

- Componente dérmico muestra maduración
- Carencia de actividad mitótica
- Infiltrado inflamatorio en banda asociado a fibroplasia o desaparición dérmica de melanocitos.

Histología

- Migración ascendente de Melanocitos.
- Retención de pigmento en estrato corneo
- Nidos grandes, simétricos, rodeados por una hendidura que lo separa de los queratinocitos
- Puentes entre las crestas hasta en un 85% de nevi acrales benignos

Nevus Acrales



Propagación Pagetoide de melanocitos en la epidermis.
Los nidos de melanocitos son irregulares y confluentes

Nevus Acrales

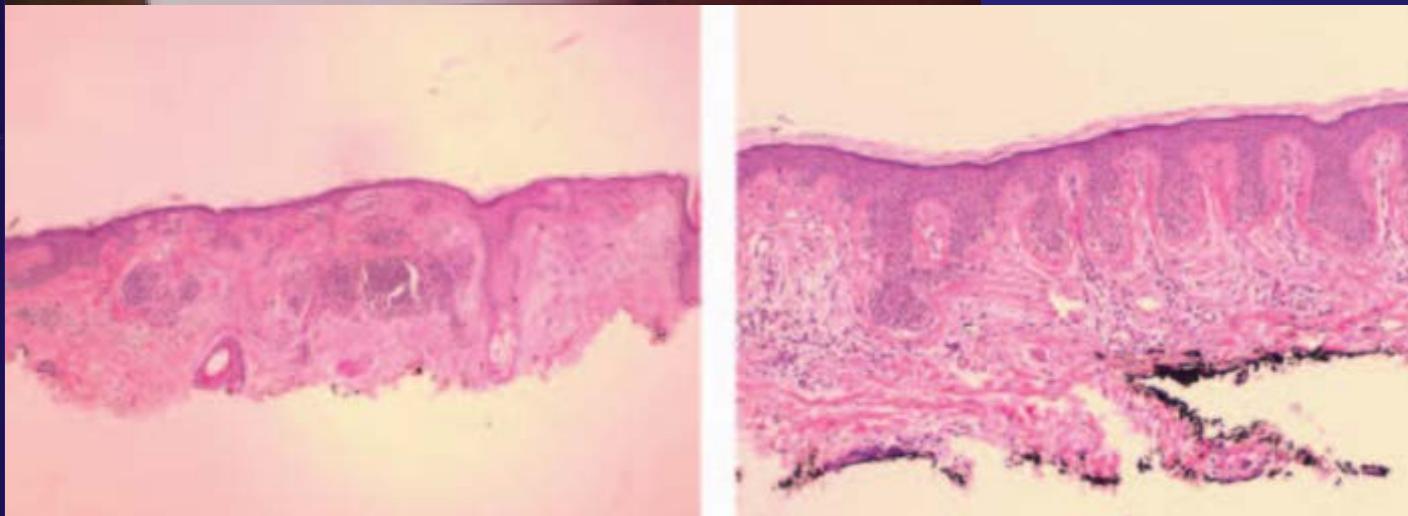
Oreja

Patrón de nidos irregulares, en forma
tamaño y localización sobre las crestas

Pobremente circunscrito

Infiltrado linfocítico, Rx estromal 81%

Nevus Acrales



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Nevus Cuero cabelludo

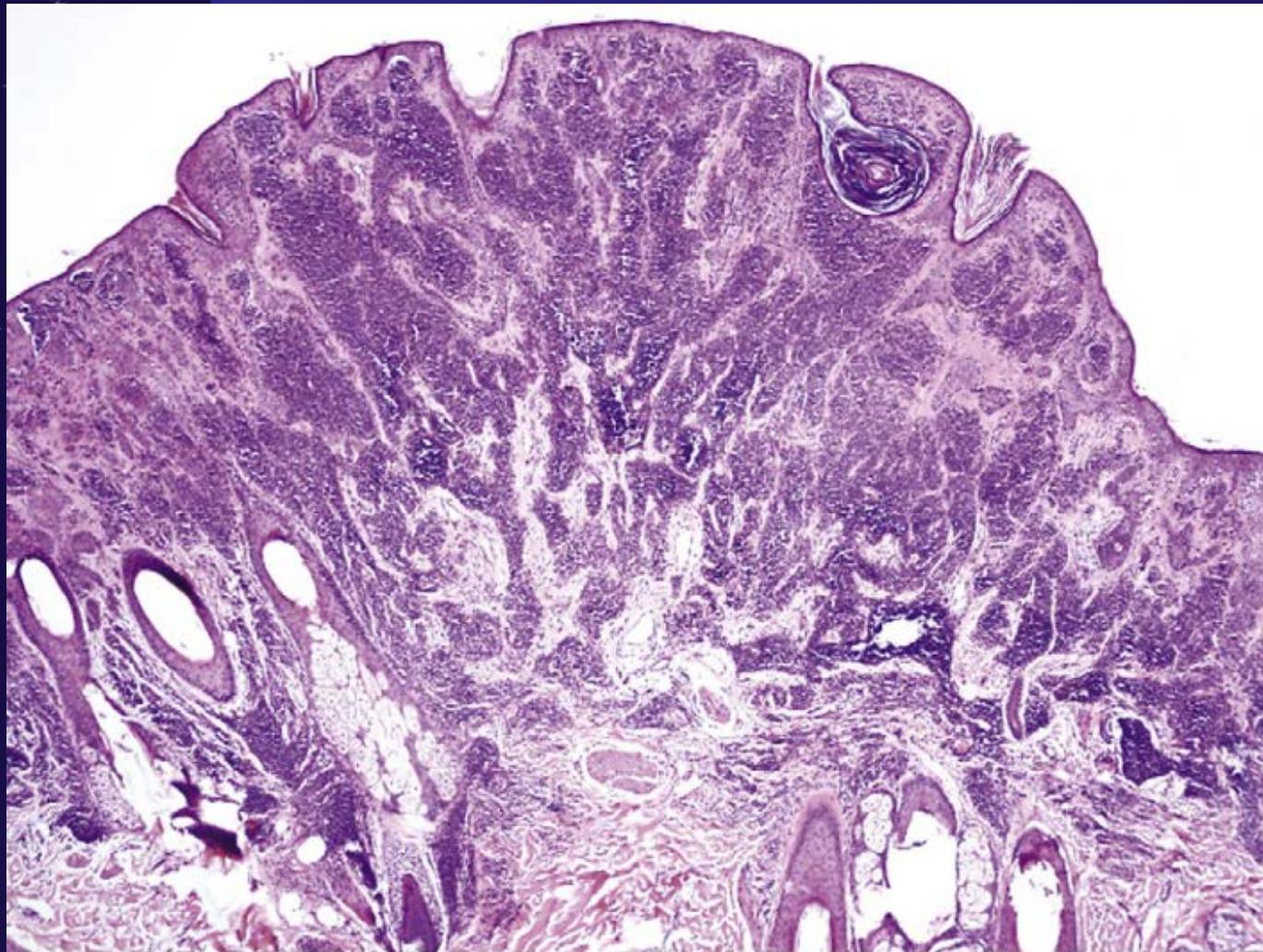
Table 2
Clinical appearance of nevi of special sites

| Special Site | Synonyms | Location | Clinical Appearance of NOSS |
|--------------|--|-------------|---|
| Scalp | <ul style="list-style-type: none">Atypical nevi of the scalp in adolescents²² | Unspecified | <ul style="list-style-type: none">Dark brownClonal nevi ("fried-egg nevi"): light brown with darker raised center^{11,32}Rim nevi ("eclipse nevi"): flat, pink, or light brown with a darker brown peripheral rim^{11,32} |

Nevus Cuero cabelludo

- Pobremente circunscritos, asimétricos
- Nidos de tamaños y formas variables (grandes)
- Diseminación a la unión dermo-epidérmica
- Base y punta de las crestas
- Extensión anexial
- *Nidos de Melanocitos grandes, dispersos a la unión dentro del folículo*

Histología



Nidos grandes de formas y tamaños irregulares en la unión dermoepidérmica y extensión perianexial.

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Histología

Table 3

Histologic comparison among nevi of special sites

| | Genitalia | Flexural Skin/Milkline | Acral Skin | Scalp (Adolescents) | | |
|------------------------------|---|---|-----------------------|---|---|---|
| | Vulva Penis ^b | Axillae, inguinal folds, umbilicus, scrotum, perianum, pubis | Breast | Ears Palms & Soles | | |
| Architectural pattern | | | | | | |
| • Circumscription | Present ⁶ | ND | Present ¹² | Minority ^{20,21} | Present ^{10,13,16,19} | Poor ²² |
| • Symmetry | Present ⁶ | ND | Present ¹² | Present ^{20,21} | Present ^{10,13} | Asymmetric ²² |
| • Nests | <ul style="list-style-type: none"> • Diminished cohesion^{6,7} • Varied size^{6,7} • Often oval in shape^{6,7} • 3 patterns: <ol style="list-style-type: none"> 1. Nested 2. Dyshesive 3. Crowded | <ul style="list-style-type: none"> • Diminished cohesion⁸ • Varied size, shape, and position at dermal-epidermal junction⁸ • Horizontal confluence of enlarged nests⁸ | ND | <ul style="list-style-type: none"> • Irregular^{20,21} • Confluent²⁰ • Located at rete tips & shoulders >> tips only²¹ | <ul style="list-style-type: none"> • Large¹⁹ • Variable size, shape^{14,19} • Positioned at dermal-epidermal junction^{14,19} | <ul style="list-style-type: none"> • Large, confluent²² • Variable in shape²² • Apparent random scattering²² • Located at rete shoulders & tips²² |
| • Lentiginous proliferation | Present ⁶ | Present ⁸ | Absent ¹² | Present ²⁰ | Present ^{14,16,19} | Present ²² |
| • Rete ridges | Bridging ⁶ | ND | ND | Elongated, bridging ²⁰ | Elongated, bridging ^{16,19} | ND |

Histología

Table 3

Histologic comparison among nevi of special sites

| Genitalia | Flexural Skin/Milkline | Breast | Acral Skin | Scalp (Adolescents) | | |
|------------------------------|---|---|-----------------------|---|---|---|
| Vulva Penis ^b | Axillae, inguinal folds, umbilicus, scrotum, perianum, pubis | Breast | Ears | Palms & Soles | | |
| Architectural pattern | | | | | | |
| • Circumscription | Present ⁶ | ND | Present ¹² | Minority ^{20,21} | Present ^{10,13,16,19} | Poor ²² |
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Histología

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| Vulva Penis ^b | Axillae, inguinal folds, umbilicus, scrotum, perianum, pubis | Breast | Ears | Palms & Soles | | |
| Architectural pattern | | | | | | |
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Histología

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Histologic comparison among nevi of special sites

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|------------------------------|--|---|---|
| Vulva Penis ^b | Axillae, inguinal folds, umbilicus, scrotum, perianum, pubis | Breast Ears | Palms & Soles |
| Architectural pattern | | | |
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| • Symmetry | Present ⁶ | ND | Present ^{10,13} |
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| • Lentiginous proliferation | Present ⁶ | Present ⁸ | Present ^{14,16,19} |
| • Rete ridges | Bridging ⁶ | ND | Elongated, bridging ²⁰ |
| | | | Elongated, bridging ¹³ |
| | | | ND |

Diagnóstico

- Difícil por ausencia de criterios diagnósticos

Diagnóstico Diferencial

- Dx: Nevus de sitio especial es ambiguo:

Los sitios especiales no están exentos de nevus displásicos y melanoma

Los N. displásicos, melanoma y N. de sitio especial comparten características

Diagnóstico Diferencial



Differential Diagnosis NEVI OF SPECIAL SITES

- Stereotypical benign melanocytic nevi
- Dysplastic nevi
- Melanomas Primario de mama <5%
- Congenital nevi
- Spitz nevi
- Traumatized nevi

Management of these melanocytic tumors is varied. Therefore, accurate diagnosis is essential.

E. Paget Pigmentada
Ca de mama metastásico
epidermotrópico pigmentado
Melanosis Benigna del Pezón

Gracias!