Coccygeal Polypoid Eccrine Nevus

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- Skin tags are fairly common lesions usually seen in adults on the neck and in body folds. The sacrococcygeal region is an unusual location for skin tags in children and may represent a congenital malformation of the spine or an isolated skin lesion. In this review, we summarize the clinical presentation, histopathologic changes, and differential diagnosis of coccygeal polypoid eccrine nevus as a rare cause of sacrococcygeal papules in children. (Arch Pathol Lab Med. 2019;143:890–892; doi: 10.5858/arpa.2018-0055-RS)

Although prevalent in adults and associated with obesity and aging, skin tags are rare in children and may warrant further evaluation. In children, the most-common locations for skin tags are the preauricular region and the vaginal region. Preauricular skin tags occur in 5 to 10 patients per 1000 live births. They may be associated with hearing impairment in neonates, and thus neonates with preauricular skin tags should be screened for hearing loss. Vaginal skin tags usually regress within a few weeks. Children with coccygeal skin tags often warrant concern because occult spinal dysraphism may be suspected; in addition to skin tags, sacral hemangiomas, dimples, or clusters of hair may also overlie a spinal malformation. Although present in the coccygeal region, coccygeal polypoid eccrine nevi, hamartomas with increased number or size of eccrine glands with no vascular proliferation, are not known to be associated with spinal dysraphism.

CLINICAL FEATURES

About 20 cases of eccrine nevi have been reported in the English-language medical literature, with various clinical manifestations. Most eccrine nevi are associated with localized hyperhidrosis, but asymptomatic lesions have also been reported. There is no predominance in gender distribution, and eccrine nevi are uncommon at birth. They are not associated with cutaneous or internal malformations. The unusual location of the coccygeal eccrine nevus as a flesh-colored papule on the midline of the coccyx (Figure 1) has been reported in 5 cases (Table 1). All but one of those lesions occurred in young girls and were present at birth. In addition, none of the reported cases was associated with internal malformation, except one with imperforate anus and unilateral multicystic kidney dysplasia.

HISTOPATHOLOGIC FINDINGS

Hematoxylin–eosin–stained sections demonstrate a polypoid fragment of skin with a slightly acanthotic epidermis with overlying hyperkeratosis (Figure 2, A). That finding may be attributed to the coccygeal location, in which frictional microtrauma occurs frequently. Within the reticular dermis, collections of well-formed eccrine ducts and glands are present. The dermal vascularity is within reference range in distribution, with no angiomatosus components surrounding the eccrine units (Figure 2, B).

DIFFERENTIAL DIAGNOSIS

Table 2 summarizes the differential diagnoses. Coccygeal polypoid eccrine nevi should be distinguished from other eccrine nevi. An eccrine nevus is a hamartoma characterized by an increase in the number or size of eccrine glands. Eccrine nevi are localized hyperhidrosis not associated with other skin abnormalities. Eccrine nevi are usually noted during childhood but are uncommon at birth. Eccrine nevi are common on upper extremities. Eccrine angiomatosus hamartomas occur predominantly on the lower extremities as a pink-red patch or thin plaque, from 1 to 10 cm in diameter. They may be associated with pain, hypertrichosis, itching, and hyperhidrosis. Microscopically, the eccrine angiomatous hamartoma is composed of prominent capillary proliferations accompanied with eccrine units.

A hair follicle nevus is a congenital nodule made up of well-formed vellus hair follicles. They are relatively rare, benign, congenital lesions and are most frequently found on the face and neck. Skin tags are generally polypoid, with a flattened epidermis overlying loose collagen bundles and an absence of adnexal structures. Vulvar skin tags, which can be seen early in life, are similarly polypoid with a connective tissue core, but there may be a few sebaceous glands attached to the epidermal surface. Eccrine units are not prominent. The adnexal polyp of neonatal skin presents as a small congenital polypoid lesion and typically is solitary. It is skin colored and regresses spontaneously within a few days of birth. It most commonly affects the areola of the breast, but other sites such as the scrotum and labia majora have been described. Pathologically, the polyp core generally includes eccrine units as well as other structures including hair follicles and sebaceous glands.
Coccygeal polypoid eccrine nevi can also be distinguished from other sacrococcygeal lesions in the pediatric age group, such as teratoma, sacrococcygeal eversion, and tailgut cysts. In pediatric patients, teratomas, a form of dermoid cyst, are the most common presacral lesion. Teratomas are mostly benign and are identified as palpable presacral or pelvic masses upon rectal examination and may extend into the abdomen. Most sacrococcygeal teratomas are seen after birth and are diagnosed before the age of 6 months.

Sacrococcygeal eversion is a condition in which the distal sacral and coccygeal vertebrae are curved in a retroverted direction. This malformation results in a tail-like protrusion that is identified by varied histologic markers, including fatty tissue, nerve fibers, striated muscle fibers, and/or cartilage. Finally, tailgut cysts are congenital malformations in the presacral space and are a malformed remnant of the hindgut. Tailgut cysts predominantly affect women, and while they can present in patients of any age, they usually appear between the ages of 30 and 60. Tailgut cysts can be asymptomatic or cause additional complications, such as a

Table 1. Review of Previously Reported Cases of Coccygeal Polypoid Eccrine Nevus

<table>
<thead>
<tr>
<th>Source, y</th>
<th>Age, y/Sex</th>
<th>Size, mm</th>
<th>Symptoms</th>
<th>Specific Adnexa</th>
<th>Presence of Other Adnexa</th>
<th>Vascular Ectasia</th>
<th>Association With Internal Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mahdavy and Smoller,13 2002</td>
<td>Birth/F</td>
<td>5</td>
<td>No</td>
<td>Proliferation of eccrine ducts and glands</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Oh et al,14 2007</td>
<td>Birth/F</td>
<td>3</td>
<td>No</td>
<td>Proliferation of eccrine glands and ducts without any angiomatous component</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Park and Lee,15 2009</td>
<td>Birth/M</td>
<td>5</td>
<td>No</td>
<td>Proliferation of eccrine glands and ducts</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Park and Lee,15 2009</td>
<td>Birth/F</td>
<td>5</td>
<td>No</td>
<td>Proliferation of eccrine glands and ducts</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Frouin et al,16 2016</td>
<td>Birth/F</td>
<td>7</td>
<td>No</td>
<td>Proliferation of eccrine sweat glands surrounded by a few healthy adipocytes</td>
<td>No</td>
<td>No</td>
<td>Yes (imperforate anus and unilateral multicystic kidney dysplasia)</td>
</tr>
</tbody>
</table>
prolapsing anal cyst, infection, or organ compression, leading to digestive or urinary dysfunction. Although the lining of tailgut cysts can include different epithelium types, including columnar, transitional, or squamous tissues, glandular or transitional epithelium differentiate tailgut cysts from epidermoid and dermoid cysts.

**CONCLUSIONS**

Coccygeal polypoid eccrine nevus is a benign unique eccrine nevus located in the coccygeal area, characterized histopathologically by normal or hyperplastic eccrine glands surrounded by normal fibroadipose tissue with occasional overlying acanthotic and hyperkeratotic epidermis. Because so few cases have been reported, the inheritance pattern is unknown, and most cases are not associated with other internal defects.

**References**


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**Table 2. Differential Diagnoses of Congenital Polypoid Eccrine Nevus**

<table>
<thead>
<tr>
<th>Disease</th>
<th>Clinical Presentation</th>
<th>Age Group</th>
<th>Primary Anatomic Location</th>
<th>Histopathologic Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sacrococcygeal teratoma</td>
<td>Palpable dermoid</td>
<td>Pediatric</td>
<td>Presacral or pelvic</td>
<td>All 3 germ cells (ie, ectoderm, mesoderm, and endoderm)</td>
</tr>
<tr>
<td>Tailgut cysts</td>
<td>Varied deepening on location</td>
<td>Any age, predominantly 30–60 y</td>
<td>Presacral</td>
<td>Columnar, transitional, or squamous tissues</td>
</tr>
<tr>
<td>Sacrococcygeal eversion</td>
<td>Prominent polypoid lesion; “human tail”</td>
<td>Congenital, neonatal</td>
<td>Lumbosacrococcygeal</td>
<td>Fatty tissue, nerve fibers, striated muscle fibers, or cartilage</td>
</tr>
<tr>
<td>Eccrine nevi</td>
<td>Linear or zosteriform papule arrangement, without overlying skin abnormalities</td>
<td>Pediatric</td>
<td>Most commonly on the forearm, also sacrococcygeal</td>
<td>Hypertrophy or hyperplasia of eccrine glands</td>
</tr>
<tr>
<td>Eccrine angiomatous hamartoma</td>
<td>Papules or nodules on extremities accompanied by hyperhidrosis and/or pain</td>
<td>Pediatric</td>
<td>Lower extremities</td>
<td>Deep dermis with increased eccrine units and capillary proliferation</td>
</tr>
<tr>
<td>Hair follicle nevus</td>
<td>Nodule on the head or neck composed of well-formed follicles</td>
<td>Congenital</td>
<td>Often head or neck</td>
<td>Epidermal hyperplasia and many hair follicles filling the dermis</td>
</tr>
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