Eyelid dermoid cyst

The term dermoid cyst does not appear to be restricted to a single kind of lesion nor is it used in only a single medical discipline. The term dermoid cyst can be found in the vocabulary of dermatologists, dermatopathologists, general pathologists, gynecologists, neurosurgeons, or pediatricians. If asked, all of these clinicians would most probably define and describe dermoid cysts differently. For example, gynecologists and general pathologists might say that a dermoid

cyst is a cystic tumor of the ovary. In contrast, neurosurgeons tend to view a dermoid cyst is associated with a congenital cyst of the spine or an intracranial congenital cyst. For pediatricians and dermatologists, dermoid cyst means subcutaneous cysts, which are usually congenital.

In all disciplines, however, the common factor is the presence of a solitary, or occasionally multiple, hamartomatous tumor. The tumor is covered by a thick dermislike wall that contains multiple sebaceous glands and almost all skin adnexa. Hairs and large amounts of fatty masses cover poorly to fully differentiated structures derived from the ectoderm.

Depending on the location of the lesion, dermoid cysts may contain substances such as nails and dental, cartilagelike, and bonelike structures. If limited to the skin or subcutaneous tissue, dermoid cysts are thin-walled tumors that contain different amounts of fatty masses; occasionally, they contain horny masses and hairs.

Pathophysiology

Dermoid cysts in the skin and subcutis occur mostly on the face, neck, or scalp.

In addition to the skin, dermoid cysts can be intracranial, intraspinal, or perispinal. Intra-abdominal cysts, such as cystic tumors of the ovary or omentum, occur as well.

Dermoid cysts have been described in persons of all ages.

- Dermoid cysts on the face, neck, or scalp are subcutaneous cysts that are usually present at birth. Intracranial or perispinal dermoid cysts are most often found in infants, children, or young adolescents.
- Intra-abdominal dermoid cysts are described in females aged 15-40 years. For example, cystic teratoma is a relatively rare tumor that most often occurs in females aged 15-40 years.

- Most dermoid cysts on the floor of the mouth occur in individuals aged 10-30 years. There are few descriptions of oral dermoid cysts in newborns or children.

History

- Dermoid cysts that are congenital and localized on the neck, head, or trunk are usually visible at birth.
- In some instances, careful medical examination may help to find most dermoid cysts.
- Intracranial, intraspinal, or intra-abdominal dermoid cysts may be suspected after specific or nonspecific neurologic or gynecologic symptoms occur. In these instances, imaging studies may help in distinguishing dermoid cysts from other tumors or organ malformations (see Imaging Studies). A congenital intracranial frontotemporal dermoid cyst may be first evident as a cutaneous fistula, although intracranial extension and cutaneous sinus tract formation are rarely seen with these dermoid cysts. [2]
- Unilateral upper eyelid swelling may be the first sign, with imaging studies demonstrating a soft tissue orbital dermoid cyst arising from the lacrimal gland. [3]
- An eyelid dermoid cyst attached to a tarsus may be evident as a firmly adherent nontender upper-eyelid nodule. [4]
- Dermoid tumors in the medial canthal area may present as masses adherent to the lacrimal canaliculi. [5]

Physical

Dermoid cysts can appear as cutaneous cysts on the head, as cysts on the floor of the mouth or elsewhere in the head, within the parotid gland,[6] or as cysts in the testes or penis. They can be quite large. [7]

Specialists in various disciplines may consider dermoid cysts to be different entities.

- Cutaneous cysts most commonly occur on the head (forehead), mainly around the eyes. Occasionally, they occur on the neck or in a midline region. When on the head, dermoid cysts are often adherent to the periosteum. The usual diameter of the lesions is 1-4 cm.
- In one study, 25 benign tumors on the forehead and brow of children and adolescents were successfully removed by means of endoscopic excision. Of these, 6 were classified as dermoid cysts. Dermoid cysts on the forehead and brow are known to cause pressure-related erosion of the underlying bony tissue, and surgical intervention may be helpful.
- In 191 children treated for congenital cysts and fistulas of the neck in 1984-1999, 21 dermoid cysts were found. Periauricular fistulas and cystic hygromas were not included in this study. [8]
- Occasionally, skin-related dermoid cysts are multiple and develop over periods as long as 20 years. In one unusual case, multiple subcutaneous dermoid cysts were present in the frontal region of a 41-year-old man, none with evident intracranial extension.
- In many patients, dermoid cysts occur on the floor of the mouth or elsewhere in the mouth. [9,10,11,12]

- Because the term dermoid is frequently used in the literature, some authors believe that this term should be used for all congenital cysts on the floor of the mouth. Three subclasses of congenital mouth cysts are described in the literature: epidermoid (simple) cysts, dermoid (complex) cysts, and teratoid (complex) cysts. Most of these lesions occur in individuals aged 10-30 years. Only a few cases describe dermoid cysts of the mouth in newborns or children.

- An unusual case of a carcinomatous transformation of a long-standing sublingual dermoid cyst has been described. [13]
- Other rare dermoid cysts in the oral cavity are those on the tongue.[14,15] As of early 2000, 17 patients with intralingual dermoid cysts are described in the English-language literature. All cases occurred in young patients. Magnetic resonance imaging (MRI) was helpful in establishing the differential diagnosis. Surgical excision corrected deglutition and speech problems in all of these patients.

- Dermoid cysts can occur elsewhere in the head.
- Dermoid cysts in the eustachian tube are rare.[16] Only 12 patients have been described. In most cases reviewed, cysts affected female patients on the left side. MRI was useful in establishing the correct diagnosis and in selecting the surgical approach.

- Reports of nasal dermoid cysts were recently published. Of 36 children with nasal dermoid sinus cysts that were treated from 1974-1994, 10 had only a midline cyst, 8 had only nasal pits, and 18 had combined cysts. Meningeal adherences have been found in only 2 patients. [17]

- Dermoid cysts can occur in the testes or penis.
- In a review of cystic testicular lesions in the pediatric population, dermoid cysts were noted. [18] Other diagnoses for these cysts include epidermoid cyst, prepubertal teratoma, juvenile granulosa cell tumor, cystic dysplasia of the rete testis, testicular cystic lymphangioma, simple cyst, and cystic degeneration after torsion. An understanding of potentially cystic testicular lesions in children leads to the best treatment choices and often to the preservation of a substantial portion of the affected testis.
- Dermoid cysts in the penis are extremely rare. Tomasini et al[3] described the first in 1997. The patient was a young white male who had significant penile swelling for several months.

- For neurosurgeons, dermoid cysts are associated with congenital cysts of the spine or intracranial cysts.

- Several cases involve ruptured cysts and generalized subarachnoid and ventricular spread of the contents (mostly fatty masses). [19,20,21]

- In some patients, spinal dermoid cysts, especially those connected to dermal sinus tract, lead to severe neurologic complications such as secondary spinal subdural abscesses caused by the spread of the infection in the dermoid cyst.

- For gynecologists and general pathologists a dermoid cyst is primarily associated with a cystic tumor of the female ovary.
- Cystic teratoma is a relatively rare tumor that most often occurs in females aged 15-40 years. A cystic teratoma consists of a thick leatherlike capsule that covers amorphous fatty masses and poorly to fully differentiated structures derived from the ectoderm. Most ovarian dermoid cysts contain skin and skin adnexa, including prominent sebaceous glands, hairs, and nails, but also teeth or eyes. Melanotic changes may also occur. Rare cases of multiple dermoid cysts of the omentum have been reported. [22] Dermoid cysts of the ovary are usually benign and easy to remove.
- Malignant melanomas may originate from melanocytes in ovarian cystic teratomas. Two new cases and 17 older cases in the literature (reported from 1903-1995) are described and were critically reviewed. The present authors found 17 additional cases of benign and malignant melanotic ovarian lesions that were not associated with a dermoid cyst, including 4 melanomas, 3 benign nevi, 5 benign melanosis, and 4 benign and malignant retinal anlage tumors. The extremely rare primary ovarian melanoma was differentiated from the more common melanoma metastatic cyst of the ovary by its unilaterality, the presence of junctional change, and detailed history taking and physical examination, the findings of which excluded other primary sites. [23]

- Three patients with metastasizing squamous cell carcinoma from a dermoid cyst of the ovary are described. [24] Malignant transformation in a dermoid cyst is a rare complication and mainly occurs in older individuals. Although the prognosis is poor, aggressive therapy may result in long-term remission.

Causes

- Dermoid cysts are true hamartomas.
- Dermoid cysts occur when skin and skin structures become trapped during fetal development.
- Histogenetically, dermoid cysts are a result of the sequestration of skin along the lines of embryonic closure

Other Problems to Be Considered

Pilar cysts, also known as trichilemmal cysts, are acquired rather than congenital. They tend to appear on the scalp rather than the face, and they tend to be intradermal rather than subcutaneous.

Although dermoid cysts are rare, they should be included in the differential diagnosis of all nodular cystlike lesions in the head or neck or in a midline (eg, chest midline) in infants and children. An intraoral nodular lesion or tumor of the tongue may be a dermoid cyst.

Cutaneous or lymph node lesions diagnosed as metastases of melanoma or squamous cell carcinoma may reflect other pathologic entities, especially in patients in whom the primary skin tumor could not been identified. Such lesions may represent a late clinical stage in the malignant malformation of melanocytic lesions, or may reflect a malignant squamous epithelial proliferation inside an ovarian dermoid cyst. Although these malignancies are extremely rare, they should be included in the dermatologic differential diagnosis.

The presence of a hair collar sign around a suspected dermoid cyst might indicate cranial dysraphism, such as that seen in a cutaneous ectopic brain.

A giant dermoid cyst of the neck can mimic a cystic hygroma, requiring MRI to differentiate

Imaging Studies

- Radiography, CT scanning, and MRI are helpful in making the correct differential diagnosis of dermoid cysts.
- MRI is particularly helpful in diagnosing intracranial or intramedullary dermoid cysts and in assessing the dissemination of fatty masses or droplets.
- MRI is helpful in planning surgical procedures and in assessing therapeutic success.
- Also see the ACR Appropriateness Criteria® suspected adnexal masses.[27]

Histologic Findings

Dermoid cysts are a result of the sequestration of the skin along the lines of embryonic closure. If connected with the ovary, dermoid cysts are true teratomas.

In contrast to epidermal inclusion cysts, dermoid cysts in the skin are lined by an epidermis that possesses various epidermal appendages. As a rule, these appendages are fully mature. Hair follicles containing hairs that project into the lumen of the cyst are often present. The dermis of dermoid cysts usually contains sebaceous glands, eccrine glands, and, in many patients, apocrine glands. Occasionally, the lining epithelium may proliferate as papillary boundaries extend externally or inward toward the lumen of the cyst. This proliferation may have some superficial resemblance to epidermal carcinomatous proliferation, and the growth may be misclassified as a cancer.

Dermoid cysts in the ovary (cystic teratomas) or those disseminated intra-abdominally may contain other structures such as nails, hairs, or cartilage and bone fragments. These cysts have cell walls that are almost identical to those of the skin, and they may contain multiple adnexal skin structures such as hair follicles, sweat glands, and occasionally, hair, teeth, or nerves.

A congenital dermoid together with a bronchogenic cyst of the tongue is extremely rare but has been described in a few patients.

Surgical Care

Surgical excision is the treatment of choice in any localization.

- Surgically remove dermoid cysts.
- In some patients, surgery should be performed even more carefully than usual because the fatty content of the cyst may spread to the surrounding tissues or anatomic structures, especially if the cyst is infected with bacteria. The spread of these contents can cause foreign body reactions and severe complications.
- Minimally invasive surgical techniques have been successful in removing dermoid cysts from difficult locations, such as those on the tongue or the floor of the mouth. Intralingual dermoid cysts lead to lingual motility defects and speech problems. These cysts should be surgically removed to restore normal lingual function and to correct speech problems.
- Intracranial, intramedullary, and ovarian dermoid cysts are difficult to treat. Sophisticated neurosurgical or gynecologic surgical techniques are often needed to remove the dermoid cyst

and prevent possible complications. High-definition fiber tracking guidance may be beneficial in the resection of an intraparenchymal dermoid cyst by means of a minimally invasive endoscopic port. [29]

- In some patients with dermoid cysts on the forehead and brow, successful excision with endoscopy-assisted surgery have been described. [30]
- In the reported cases, no complications (eg, paresthesia or numbness on the scalp) occurred.
- The absence of visible scarring is an additional advantage of endoscopy-assisted surgery.

- Angular dermoid cyst excision using an eyelid crease approach may yield excellent cosmesis

Complications

- Several possible complications of spontaneous or posttraumatic rupture and surgical procedures have been described.
- In patients with a ruptured spinal dermoid cyst, fatty droplets can disseminate in the cerebrospinal fluid or in a dilated central canal of the spinal cord.
- In other patients, subarachnoid and ventricular fat dissemination can occur after the cerebellopontine angle dermoid cyst is resected.
- Spinal subdural abscesses are a possible complication because of the bacterial infection of spinal dermoid cysts in a dermal sinus tract.
- A ruptured intracranial dermoid cyst be an incidental finding on an MRI performed for other purposes or because of a persistent headache. [32]

- Pay special attention to intralingual dermoid cysts because deglutition and speech problems may occur.
- Malignant transformation is an unusual complication that may occur in patients with long-standing dermoid cysts.
- Carcinomatous transformation to a squamous cell carcinoma is described in sublingual and intra-abdominal dermoid cysts, most often dermoid cysts of the ovary
- Metastatic malignant melanomas arising from dermoid cysts have been reported in the literature.

28