Pathology of the Ear

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ABSTRACT

The external ear is exposed to weathering and trauma; it also has sparse vascularity, making it prone to infection and disease. The external location of the cutaneous ear makes it easily visible for diagnosis and accessible for treatment. In this article, the authors focus on diseases of the ear that are most commonly encountered and may be subject to surgical and medical evaluation and/or treatment. Epidemiology, pathogenesis, clinical course, and treatment for each disease entity are discussed.

KEYWORDS: Pathology, external, ear, dermatology, surgery

The ear is an important organ that is crucial for translation of air vibrations into sound. It also plays a lead role in the sense of balance and position of the body. Its exterior location allows clinicians to diagnose cutaneous disease with ease if equipped with the appropriate knowledge of pathologic conditions. In this article, we focus on the diseases of the external ear that are commonly encountered, and the medical and surgical evaluation and treatment of these diseases. Diseases are classified into morphologic groups.

ANATOMY

The external ear consists of two biologically significant sections used to capture the air vibrations, which the inner ear converts into audio stimulus. The external section is called the auricle and is composed of a single thin plate of elastic cartilage covered with a thin layer of skin, which contains sebaceous glands as well as fine hairs. The cartilage is composed of numerous elastic fibers and is known for its ability to tolerate distortion without damage.

The auricle is an oval-shaped structure and is directed slightly forward with a concave surface. Many

defined ridges and valleys shape the lateral surface of the auricle. The prominent outer ridge is called the helix, which gives the overall shape of the ear. The helix curves down and connects to the lobe, which is the inferior portion of the auricle. The lobe contains no cartilage and is composed of tough areolar and adipose tissue, making it the most vascularized portion of the external ear. The antihelix is also a curved ridge, which lies interior and parallel to the helix. The antihelix splits and forms a “Y” as it approaches the superior portion of the auricle; this split forms the fossa triangularis. The valley between the helix and antihelix is called the scapha. The deepest depression, which leads directly to the external auditory canal, or acoustic meatus, is called the concha. The tragus (superior) and antitragus (inferior) are separated by the intertragic notch located in front of the concha and mark the beginning of the external auditory canal.

The external auditory canal is the second portion of the external ear. This tunnel is a passageway into the temporal lobe of the skull, which extends inward and slightly upwards and comes to an end at the tympanic membrane. The outer segment of the canal is lined with a membrane-containing ceruminous glands and small

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protective hairs. The inner segment does not contain any specialized cells.

FLESH-COLORED ANOMALIES

Papules

ACCESSORY TRAGUS

Epidemiology: Accessory tragus is a relatively common congenital anomaly seen in ~3 to 6 per 1000 live births.¹ Familial occurrences are sometimes seen.

Pathogenesis: Accessory tragi are formed during development from remnants of and/or extra tubercles of the first branchial arch.

Clinical Course: These lesions typically manifest as skin-colored papules or nodules in the preauricular area, rarely on the mandibular cheek or neck. Although most are solitary, they may be multiple and are bilateral in ~10% of cases.¹

Treatment: Infants with isolated accessory tragi should have an assessment of their hearing because they are at increased risk for hearing impairment.² Careful evaluation for other dysmorphic features is necessary as this malformation may be part of multiple congenital anomaly syndromes.² Surgical excision is curative and special care must be taken to ensure removal of any protruding cartilage.

GOUTY TOPHUS

Epidemiology: The National Health and Nutrition Examination Survey III showed that the overall prevalence of gout was 2% in men older than 30 and women older than 50.³ Chronic tophaceous gout manifests as dispositions of monosodium urate crystals in the soft tissue and the ear is the most common location for tophi to form in the head and neck region.

Pathogenesis: The underlying mechanism of gout is altered purine metabolism leading to hyperuricemia. Local solubility limits of uric acid are exceeded; and monosodium urate crystal deposition in joints and soft tissues lead to the clinical manifestations of acute gouty arthritis and chronic tophaceous gout. Commonly reported modifiable risk factors include a high-purine diet, alcohol use, obesity, and diuretic therapy.

Clinical Course: Over time, frequent attacks of acute gout can lead to chronic tophaceous gout. Chronic tophaceous gout usually presents on average ~10 years after the initial onset of gout. Auricular tophi present clinically as firm dermal or subcutaneous papules/nodules. They may be smooth or multilobulated and flesh-colored or whitish yellow with surrounding erythema. Tophi may ulcerate through the skin and protrude a chalky yellowish-white material.

Treatment: Urate-lowering therapy is necessary for chronic tophaceous gout. Allopurinol is first-line

treatment in patients without renal disease. Uricosuric agents like Probenecid are second-line treatment for patients intolerant of allopurinol or patients with refractory disease. Febuxostat, a nonpurine xanthine oxidase antagonist, is an investigational drug not yet approved by the U.S. Food and Drug Administration that in recent studies was shown to be comparable to allopurinol in lowering uric acid levels.

NONMELANOMA SKIN CANCER

Epidemiology: It is estimated that two million cases of nonmelanoma skin cancer were diagnosed in the United States in 2004 with the predominance being basal cell carcinoma (BCC). However, incidence reports of BCC are imprecise because there is no cancer registry that collects data on BCC.¹ Noteworthy, 60 to 80% of all BCC occur in the head and neck region.¹ The ear is a unique location because certain anatomic points on the ear favor one type of nonmelanoma skin cancer over the other. In the pinna, basal cell predominates; however, in the external auditory canal, squamous cell carcinoma (SCC) is more common.⁴ Both types tend to have a male predominance and are also found more commonly in Caucasians, people over the age of 40, and people living in higher geographic latitudes.

Pathogenesis: BCC arises from malignant transformation of keratinocytes in the basal layer of the epidermis. A well-known pathogenic mechanism for the development of BCC involves the tumor suppressor gene, patched 1 (PTCH1), which is thought to become inactivated in a two-hit model like that of the retinoblastoma gene.¹ Like the other types of skin cancers, both environmental and genetic factors play a role in the development of BCC. Associated risk factors for nonmelanoma skin cancers are directly related to a person's overall sun exposure or susceptibility to solar radiation including environmental and occupational sun exposures, having fair skin that easily burns, or having an inherited condition of increased susceptibility to UV radiation. Additional factors include chronic arsenic exposure, therapeutic exposure to photochemotherapy (PUVA; especially for SCC), smoking, exposure to ionizing radiation, chronic infection with human papillomavirus, immunosuppression, and chronic nonhealing wounds.¹

Clinical Course: Anatomically speaking, nonmelanoma skin cancer of the ear presents differently in different regions. The pinna of the ear parallels the skin cancer presentation typically seen in other areas of the body, with basal cell being more frequent.⁴ However, the external auditory canal of the ear has a higher frequency of squamous cell, and in this region it mimics otitis externa with a history of chronically draining otorrhea or otalgia.⁴ Nodular BCC is the most common subtype and classically presents as a pink or flesh-colored nodule with rolled borders and telangiectasias. However,

BCC can be pigmented and present as clinically similar to melanoma. BCC of the auricle is considered a high-risk lesion because it arises in the H-zone of the face, an area representing embryologic fusion planes where there is little resistance to invasion by the cancer. Although basal cell carcinomas can be locally destructive, metastasis is extremely rare. In contrast, SCC is much more likely to metastasize. SCC can have a variable presentation depending on whether it is well or poorly differentiated, ranging from a firm, erythematous, hyperkeratotic papule or nodule to a soft, fleshy, granulomatous papule or nodule with or without keratosis (Fig. 1). SCC often presents at a later stage than BCC and tends to recur locally with less-invasive treatment.

Treatment: Periauricular basal cell carcinomas are treated primarily by Mohs micrographic surgery for two main reasons: their location places them in a high-risk category and good cosmetic reconstruction in this area is necessary to retain structure and function of this important anatomic site. Occasionally, superficial BCC can be treated with electrodesiccation or topical imiquimod. For SCC of the external ear canal, a more aggressive approach may be necessary based on the depth of invasion and may include resection of the tumor with temporal bone resection and/or lymph node dissection.⁴

Plaque

ACANTHOMA FISSURATUM

Epidemiology: Acanthoma fissuratum occurs in the postauricular sulcus or upper lateral nose at the site of chronic low-grade pressure or rubbing, especially in those with ill-fitting eyeglass frames. Exact epidemiology is not available, as many of these patients never seek help.

Pathogenesis: Pressure and rubbing by the eyeglass frame leads to chronic irritation and collagen deposition. As the body tries to eliminate the abnormal collagen, local inflammation ensues.¹

Figure 1 Squamous cell carcinoma.

Clinical Course: Clinically, there is a skin-colored to erythematous plaque or nodule in the upper postauricular sulcus with a central vertical depression where the eyeglass stem lies.

Treatment: Within months of discontinuing, replacing, or properly adjusting eyeglass frames, the lesion should resolve. If the lesion persists, excision should be considered to rule out other diagnoses.

Nodules

CHONDRODERMATITIS NODULARIS HELICIS

Epidemiology: Chondrodermatitis nodularis helices (CNH) is a common condition in clinical practice, but is rarely reported in the literature, as evidenced by only 600 cases that have been reported between 1966 and 2004.⁵ It affects elderly men and women equally, but the location of the lesion has gender specificity, with the antihelix favoring women and the helix favoring men.¹

Pathogenesis: The exact cause is unknown, but factors frequently implicated in the development of lesions include solar damage, cold, trauma, local ischemia, and pressure. It is thought that these lesions may arise from pressure (hearing aids, sleeping on affected side, phone ear pieces, and hats) or inflammation that impedes the vascular supply of the cartilage thereby leading to subsequent degeneration, remodeling, and extrusion of the collagen.^{5,6} Autoimmune mechanisms have also been suggested as there have been reports of autoantibodies to type II collagen.⁵

Clinical Course: Clinically, the lesions present as chronic, tender nodules of the ear with either a central keratin-filled crater or an active sinus tract. The nodules usually start out less than 10 mm in size and then rapidly enlarge to a maximum size before remaining stable. They can appear pink to pearly gray in color. This presentation is secondary to the underlying degenerative changes in the collagen, which produce the extrusion-like changes as described above.

Treatment: Given the benign nature of this condition, conservative management is the most practical treatment. A simple approach would be avoidance of the triggering factors, such as sun exposure or pressure from various objects. Intralesional steroid injections and collagen implants are other more conservative approaches. More-invasive surgical options include excision of the cartilage alone or wedge resection, the latter of which is preferred by otolaryngologists, but may result in less-appealing cosmetic results.⁵ Cryosurgery, electrodesiccation and curettage, and CO₂ laser therapy are other alternatives.

KELOID

Epidemiology: Keloids are more common in darkly pigmented skin and incidence rates have been reported as

high as 16% in some African American populations.¹ Women and men are equally affected and there is a familial tendency toward developing these lesions.

Pathogenesis: Keloids are considered to be a deviation from the normal healing process and are triggered by numerous inflammatory stimuli. They can be precipitated by trauma, but can also develop without any apparent injury. Events known to precede auricular keloids include ear piercing most commonly; however, keloids of the auricle also occur after otoplasty or infection secondary to nickel allergy.

Clinical Course: Keloids classically present as dense, firm dermal scar tissue that is raised above the skin as a purplish nodule. Keloids are known to extend beyond the boundaries of the original wound into adjacent tissue and rarely regress without treatment. They manifest clinically by pain or pruritus, although the unsatisfactory cosmetic appearance may be the presenting complaint.

Treatment: Small keloids on the earlobe can be treated with simple excision although larger lesions may require a split-thickness skin graft. The risks of further trauma and iatrogenic keloid formation must be weighed against the potential benefits of surgery. Recurrence rates after surgical excision alone can be very high. Adjuvant approaches include silicon occlusive dressings, mechanical compression, radiation, cryosurgery, topical imiquimod application, bleomycin tattooing, intralesional injections of steroids, 5-fluorouracil, as well as interferons.

SUBEPIDERMAL CALCIFIED NODULE

Epidemiology: Subepidermal calcified nodules are often found in children but have also been reported in infants and adults.

Pathogenesis: Pathogenesis is unknown; however, it has been hypothesized that trauma,⁷ perhaps in utero, or calcification of preexisting milia, nevi, or eccrine duct hamartoma may be the cause.¹

Clinical Course: It presents most often as a firm, whitish, verrucous papule or nodule, usually with erythema. It is typically found on the head and neck, especially the ears, but is also seen on the lateral aspects of the digits.

Treatment: If lesions are symptomatic, surgical removal is the treatment of choice.

Cysts

AURICULAR PSEUDOCYST

Epidemiology: Auricular pseudocyst occurs most commonly in middle-aged men.

Pathogenesis: The pathogenesis is unknown; however, it has been suggested that cavity formation during embryogenesis or repeated trauma or mechanical stimulation are possible etiologies.^{1,8}

Clinical Course: The lesions are usually unilateral and present as a painless, fluctuant swelling that develops over a period of weeks. They measure ~1 cm in diameter and are observed most commonly in the scaphoid fossa of the ear.⁹

Treatment: Treatment includes aspiration, with or without coadministration or treatment alone with intralesional corticosteroids or sclerosing agents. Incision and drainage with destruction of the cavity is another option. All treatments should be followed by application of a pressure dressing.

EPIDERMOID CYST

Epidemiology: Epidermoid cysts are the most common cutaneous cysts and usually occur in young to middle-aged adults.

Pathogenesis: Cystic enclosure of the epithelium within the dermis causes an enclosure that fills with keratin and lipid-rich debris.

Clinical Course: It usually presents as one or a few dermal nodules, ranging in size from 0.5 to 5 cm.¹⁰ A visible central punctum may be seen, representing the follicle from which the cyst is derived. They occur most commonly on the face, scalp, neck, and trunk; upon pressure they may express a keratinaceous material, often with an unpleasant odor. Rupture of the cyst wall may lead to a painful inflammatory reaction.

Treatment: Surgical excision is curative if the entire cyst wall is removed. Inflamed epidermoid cysts may require incision and drainage, antibiotic therapy, or intralesional triamcinolone.¹

ERYTHEMATOUS APPEARANCE

Macule

ACTINIC KERATOSIS

Epidemiology: Actinic keratoses (AK) are seen most commonly in those with sun-damaged skin and become progressively more common in middle age. Males are more commonly affected and multiple lesions are often seen in skin types I and II.

Pathogenesis: Inadequate sun protection and chronic exposure to sunlight, especially ultraviolet B (UVB) rays leads to cumulative damage of keratinocytes. It is estimated that anywhere from 10 to 20% of these lesions may develop into squamous cell carcinoma in one or more lesions.¹ p53 mutations may be involved in the progression of actinic keratoses to squamous cell carcinomas.¹¹

Clinical Course: Actinic keratoses arise on sun-damaged skin, especially on the tops of the ears, upper forehead, scalp, nasal bridge, malar eminences, dorsal hands, and extensor forearms. Lesions typically range in size from a few millimeters to several centimeters in

diameter and usually present as poorly defined areas of erythema with overlying scale. A clue to their presence is underlying dyspigmentation, telangiectasia, and wrinkling, indicating a history of solar damage.

Treatment: The best treatment is prevention by means of sun avoidance with sun-protective clothing and sunscreens. The most commonly used treatment modality is cryosurgery; however, other options are frequently implemented in those with significant photodamage, multiple, or recurrent lesions and include 5-fluorouracil, imiquimod, topical and systemic retinoids, facial peels, laser surgery, and photodynamic therapy.^{1,9}

Plaques

DISCOID LUPUS ERYTHEMATOUS

Epidemiology: Cutaneous lupus erythematosus encompasses a broad range of clinical appearances, this section will focus on discoid lupus erythematosus (DLE), a subdivision under lupus erythematosus-specific diseases that is known to manifest on the ears and is also the most common cutaneous manifestation of lupus. DLE is a common disease that displays a remarkable female predilection. Ethnicity is also a major risk factor toward the development of lupus, particularly DLE, which is known to affect African Americans more than Caucasians.

Pathogenesis: Systemic lupus erythematosus and all of the subtypes are known to be autoimmune in nature, as evidenced by the numerous autoantibodies that have been documented in patients, including smith (Sm), nucleosomes, histones, and double-stranded (ds) DNA. However, DLE is distinguished by having a high titer of ANA in ~5% of patients, generally no anti-dsDNA antibodies and rarely antibodies to Ro/SSA or U1-RNP. A proposed mechanism for the development of this autoimmunity includes an initial infectious agent that cross-reacts with self-antigen in a genetic background favorable to self-antigen reactivity.¹

Clinical Course: Distinguishing characteristics of DLE lesions include sharply bordered erythematous, keratotic plaques that show a coin-shaped appearance. Areas of predilection of the localized form of DLE include the face and scalp, especially the cheeks, forehead, ears, nose, and upper lip. The centers of the lesions are described as having firmly adhered areas of white, follicular hyperkeratosis that are exquisitely painful if lifted. Over time, these plaques become atrophic and scar or produce alopecia in hair-bearing areas. Involvement of the auricles can lead to significant disfigurement.

Treatment: The reported treatment options available for DLE are extensive. Consistent photoprotection is emphasized. Corticosteroids are the mainstay of topical treatment in cutaneous lupus erythematosus, including DLE.

LUPUS PERNIO (SARCOIDOSIS)

Epidemiology: Sarcoidosis occurs in patients of all races, ages, and sexes, but is most prevalent between the ages 25 to 35 years and 45 to 65 years, which demonstrates a bimodal distribution. However, the prototypical sarcoid patient is an African American female, given this is the group with the highest incidence, reported to occur at a rate of 107 per 10,000.¹ Cutaneous manifestations of sarcoidosis are common, with incidence reported between 9 to 37%.¹² In this section, we will discuss lupus pernio, a distinctive subset of cutaneous sarcoidosis that manifests in our region of interest, the auricle.

Pathogenesis: Sarcoidosis is a multisystem granulomatous disease. Initiating factors include hyperactivity of the cell-mediated immune system with upregulation of CD4⁺ T-helper cells and subsequent formation of epithelioid granulomas in a variety of tissues. The antigen-triggering granuloma formation in patients with sarcoidosis is unclear. Some theories include an autoimmune etiology or infectious cause. Genetic susceptibility is demonstrated by the association of sarcoidosis with HLA-1, HLA-B8, and HLA-DR3.¹ Environmental and occupational associations have also been reported.

Clinical Course: Lupus pernio presents as violaceous papules and nodules that coalesce to form plaques and are found primarily on the nose, cheeks, and ears. The lesions are fibrotic and are known to scar and distort the structures on which they appear, including the auricle and any adjacent bone. Lupus pernio is associated with other fibrotic manifestations of sarcoidosis, including lung disease and bone cysts. In patients with lung disease, resolution of chest x-ray abnormalities is less likely to occur in patients with concomitant lupus pernio. A possible variant of lupus pernio, termed angiolupoid lesions, is differentiated by prominent telangiectasias.¹²

Treatment: Because lupus pernio has a tendency to scar, early treatment is warranted to avoid this complication. Systemic corticosteroids are recommended as first-line agents with infliximab and antimalarials representing second- and third-line treatments.¹²

SEBORRHEIC DERMATITIS

Epidemiology: Seborrheic dermatitis is a relatively common condition, affecting 1 to 3% of the nonimmunosuppressed population.¹³ It is more common in certain comorbid conditions, such as acquired immunodeficiency syndrome (AIDS), Parkinson disease, chronic alcoholic pancreatitis, hepatitis C, and cancer.¹³ It is known to have a male predominance.

Pathogenesis: Though the mechanism is not completely elucidated, the *Malassezia* yeast species (formerly known as *Pityrosporum*) is generally accepted to play a central role in the pathogenesis of seborrheic dermatitis. Recent theories relate yeast density and/or specific species to susceptibility in different individuals. Various

endogenous host factors may explain increased susceptibility in certain groups, such as hormonal influences, immunologic, nutritional, environmental, and lifestyle factors.

Clinical Course: Seborrheic dermatitis commonly presents as erythematous patches with indistinct margins and a yellow, greasy scale. The external ears are a site of frequent occurrence, including the postauricular region and conchal bowls. Other areas affected are the sebaceous areas of the skin including the scalp, eyebrows, eyelids, nasolabial crease, chest, and back.

Treatment: There is a diversity of treatments for seborrheic dermatitis ranging from topical azoles, keratolytics and steroids all the way to oral azoles utilized in widespread or refractory disease. However, patients must be educated that treatment is not intended to be a cure; it is only meant to alleviate symptoms. For lesions involving the ears, over-the-counter shampoos containing pyridithione zinc, selenium sulfide, and ketoconazole are most popular. Prescription strength versions of these shampoos are available for persistent lesions.

Nodule

RELAPSING POLYCHONDritis

Epidemiology: Relapsing polychondritis (RP) is a rare rheumatologic condition most common in Caucasians between the ages of 20 and 60 years. About 30% of those affected have concomitant rheumatologic or autoimmune disease, such as rheumatoid arthritis, systemic lupus erythematosus, or Sjögren syndrome.¹⁴

Pathogenesis: The exact mechanism of pathogenesis remains unclear, but antibodies to type II collagen are not uncommon in affected individuals and titers may correlate with disease activity.¹⁴ However, these antibodies have low specificity given the fact that these antibodies are also found in several other rheumatologic and autoimmune diseases. A role for the humoral immune response has been suggested in relation to the cartilage matrix protein, Matrilin-1.¹ Relapsing polychondritis is also associated with HLA-DR4.¹

Clinical Course: Relapsing polychondritis is not a benign disease; approximately one quarter of patients with RP die of their disease after an average of 5 to 7 years.¹⁴ Auricular chondritis is the presenting symptom in nearly 26% of patients, while 89% will eventually develop this symptom.¹⁴ Most cases are bilateral and are characterized by erythema, pain, warmth, and swelling of the cartilaginous components of the auricle. The disease is speckled with acute episodes lasting from days to several weeks. Complications include damage to auricular cartilage resulting in a floppy, droopy ear or a cauliflower-shaped ear. These symptoms may occur on top of a background of systemic disease consisting of arthritis, respiratory

involvement, ocular manifestations, and more rarely, hearing loss, valvular heart disease, renal dysfunction, and neurologic sequelae.¹ The course of the disease ranges from a low-grade, mild condition to rapidly progressive disease. Causes of death are usually secondary to systemic complications. Spontaneous remissions are common.

Treatment: Relapsing polychondritis is a treatable disease and survival rates of up to 94% at 8 years have been reported.¹ Corticosteroids are the standard of treatment, improving acute flare-ups and decreasing the frequency and severity of recurrences, but are not considered to affect the long-term progression of the disease. Nonsteroidal anti-inflammatory drugs (NSAIDs), dapsone, and colchicine are also used. Other immunosuppressives may be used as an adjunct to steroids. Most recently, successful treatment was accomplished with infliximab.¹⁵ Surgical intervention may be required for systemic complications.

BLUE-COLORED ANOMALIES

Macule

OCHRONOSIS

Epidemiology: Ochronosis is an inborn error of metabolism; dermatologic features are rarely seen before 10 to 15 years of age.¹

Pathogenesis: Ochronosis is classically associated with alkaptonuria, a rare, autosomal recessive disease caused by deficiency of homogentisic acid oxidase, leading to accumulation of homogentisic acid in virtually all collagen-containing structures. Hydroquinone can cause an exogenous ochronosis.

Clinical Course: Homogentisic acid is an insoluble pigment that binds to collagen bundles or cartilage and causes a blue-gray pigmentation in the helices of the ear and sclerae. Abnormal pigmentation may first be seen in the axillary area; later, it may affect the entire face as well as the palmar and plantar surfaces.

Treatment: There is no definitive treatment, but administration of an inhibitor of homogentisic acid production, nitisinone, may be beneficial.^{1,16} Discontinuation of hydroquinone may lead to fading of the discoloration in exogenous ochronosis.

Papule

VENOUS LAKE

Epidemiology: Venous lakes most commonly occur on the face, lips, and ears of patients over the age of 50.

Pathogenesis: Pathogenesis is unknown, but it is thought that it may be related to solar exposure because most patients have underlying sun-damaged skin.

Figure 2 Venous lake.

Clinical Course: It presents as a small, dark blue to violaceous, soft papule (Fig. 2). Partially compressing the lesion results in drainage of most of the blood contents, helping to distinguish it from nodular melanoma.⁹

Treatment: Lesions are benign, however, may be treated for cosmetic reasons by electrosurgery, cryotherapy, hemoglobin-targeting laser, or rarely, surgical excision.^{1,9,10}

BROWN-COLORED ANOMALIES

Macules

LENTIGO MALIGNA

Epidemiology: Lentigo maligna (LM), otherwise known as melanoma in situ, accounts for up to 15% of primary cutaneous melanomas¹ and is diagnosed most frequently in the seventh decade of life. There is equal incidence between males and females.

Pathogenesis: LM occurs most often on sun-exposed areas of fair-skinned individuals and in those with chronically sun-damaged skin. If left untreated, ~5% may become invasive.^{17,18}

Clinical Course: LM is a slow growing, asymmetric flat brown to black macule. There may be distinctive variations in the hues of brown and a black-speckled pattern often overlies the lesion. It is usually well defined, but some areas may have blurred or irregular borders containing notching and “geographic” shapes with inlets and peninsulas.¹⁰ Lesions are usually 3 cm or larger and can grow up to 20 cm. Wood’s light often reveals irregular pigmentation extending well beyond the clinical lesion. The clinical change that indicates transition from LM to lentigo maligna melanoma (LMM) is the appearance of an abnormal color pattern and of papules, plaques, or nodules within the lesion.

Treatment: Treatment options are vast and include conventional surgery, micrographic Mohs surgery,

cryosurgery, radiotherapy, electrodesiccation and curettage, 5-fluorouracil, imiquimod, azelaic acid, retinoic acid, and lasers. The lowest recurrence rates, on the order of 4 to 5%, occur with micrographic Mohs surgery.^{19,20} Conventional surgery, cryotherapy, and radiotherapy also yield good results with recurrence rates of 7 to 10%.^{19,20}

JUNCTIONAL NEVI

Epidemiology: The prevalence of acquired melanocytic nevi is related to age, race, and perhaps genetic and environmental factors. Many begin to appear throughout childhood and peak in the third decade of life. With increasing age, there is gradual involution and fibrosis of the lesions and most disappear by the seventh decade.^{1,10}

Pathogenesis: Junctional nevi are hypothesized to arise from the proliferation of slightly altered melanocytes within the epidermis.¹

Clinical Course: Lesions are less than 1 cm, round or oval, medium to dark brown macules that are well circumscribed. Skin markings are preserved on the surface of the nevus. There may be some asymmetry; however, the borders are usually regular and well defined.

Treatment: In general, no treatment is needed. However, surgical excision is done in those cases in which there is a changing lesion or an atypical clinical appearance for cosmetic reasons or repeated irritation.

SOLAR LENTIGO

Epidemiology: Solar lentigines are seen most commonly on the sun-exposed areas of older individuals or in younger individuals with acute or chronic sun exposure. They are seen in 90% of the Caucasian population older than 60 years.¹ Lesions are most common in Caucasians, but also occur in Asians.^{1,10}

Pathogenesis: Solar lentigines develop in response to ultraviolet (UV) radiation exposure and are characterized by epidermal hyperplasia with variable proliferation of melanocytes and a build-up of melanin in keratinocytes.

Clinical Course: Lesions are usually multiple and appear on sun-exposed skin as 3 mm to 2 cm, well-circumscribed, round or oval macules that vary in color on a spectrum from tan to black.

Treatment: Preventative measures such as sun avoidance and sunscreens are the best treatment. Cryosurgery and laser surgery are also acceptable treatments; however, may result in post-treatment dyspigmentation. The use of bleaching creams and retinoids has been shown successful by some and ineffective by others.^{1,21} Although solar lentigines are benign, they indicate chronic UV exposure and patients should be regularly monitored for development of nonmelanoma and melanoma skin cancer.

Papules

COMPOUND NEVI

Epidemiology: The prevalence of acquired melanocytic nevi is related to age, race, and perhaps genetic and environmental factors. As mentioned above, many nevi begin to appear throughout childhood, reach a peak in the third decade and resolve by the seventh decade.^{1,10}

Pathogenesis: Compound nevi are thought to occur from the proliferation of slightly altered melanocytes within the epidermis and dermis.¹

Clinical Course: Compound nevi appear as less than 1 cm, round or oval, brown lesions with variable degrees of elevation and a smooth or cobblestone-like surface. Pigment tends to be located more centrally than peripherally.

Treatment: In general, no treatment is needed. However, surgical excision is done in those cases in which there is a changing lesion or an atypical clinical appearance for cosmetic reasons or repeated irritation.

SEBORRHEIC KERATOSIS

Epidemiology: Seborrheic keratosis (SK) is one of the most common benign tumors of the external ear and these lesions are ubiquitous in older adults. Caucasians are affected most commonly and there is a familial predisposition toward the development of these lesions. Women and men are equally affected.

Pathogenesis: Commonly reported etiologies include UV light exposure, human papillomavirus, hereditary factors, hormonal factors, and most recently, mutations in the fibroblast growth factor receptor-3.¹

Clinical Course: On the ear, they commonly present as variably pigmented waxy papules and plaques with a verrucous surface that appears stuck-on (Fig. 3). The lesion may enlarge with age and in some cases can affect the entire ear. Seborrheic keratosis is benign and very rarely acquires malignant changes, although some

Figure 3 Seborrheic keratosis.

subtypes can be confused with other malignant skin conditions.

Treatment: Treatment options include cryotherapy, electrodesiccation, simple curettage, and shave excision. Because these lesions may be confused with malignant melanoma or squamous cell carcinoma, histologic evaluation is often essential.

BLACK-COLORED ANOMALIES

Macule

MELANOMA

Epidemiology: Malignant melanoma is the third most common malignancy of the external ear, with the overall annual incidence of melanoma reaching 3 to 7%.^{1,4} Of the 20% of melanomas that arise in the head and neck, 7 to 15% arise specifically in the auricular region.⁴ Auricular melanoma is rare in children and has a predominance for men in their late 50s.⁴ Although incidence rates of melanoma continue to rise, the mortality rate has slowed due to improved detection methods.

Pathogenesis: A combination of genetic and environmental factors plays a role in the development of malignant melanoma. A history of intense, intermittent exposures to sun or an increased number of lifetime sunburns predisposes an individual to develop melanoma as does living in a higher geographic latitude. Contributing genetic factors include a personal or family history of dysplastic nevi or melanoma, inherited DNA repair defects, or the presence of a fair complexion with a tendency to burn. Both UVA and UVB are implicated in the development of melanoma, although their relationship is not as direct as it is for nonmelanoma skin cancers.

Clinical Course: Melanoma of the auricle can have a morphologic appearance like melanoma anywhere else; however, the most common subtypes arising on the ear are superficial spreading and nodular melanoma.⁴ Superficial spreading melanoma typically presents as a macular lesion with an indistinct border and variable pigmentation.¹ It proceeds through a slow radial growth phase before progressing to a rapid vertical growth phase, which is evident clinically by the presence of a papule or nodule. Because the ear is deficient in subcutaneous tissue, this anatomic site allows easier invasion into deeper tissue. Nodular melanoma bypasses the radial growth phase and presents as a nodule with variable pigmentation, which can progress to ulceration or bleeding. Initially, metastases spread to regional lymph nodes, involving the preauricular, parotid, and upper jugular nodes whereas hematogenous spread occurs later in the disease course.⁴ Prognosis depends on the stage at diagnosis, with Breslow thickness (depth of lesion) being particularly important.

Treatment: The primary treatment of malignant melanoma of the auricle is surgical, involving wide excision of the skin of the pinna.⁴ However, excision into the cartilage of the ear is optional depending on the extent of the disease, Breslow thickness, and the amount of reconstruction needed. For tumors involving the external auditory canal, wide excision may involve sleeve resection of the external auditory canal with subsequent reconstruction using a split-thickness skin graft.⁴ Sentinel node biopsy in patients with auricular melanoma is a debated topic, but most physicians agree that a lesion greater than 1 mm deep merits the procedure.⁴ Adjunctive radiation, chemotherapy, or immunotherapy is reserved for metastatic disease.

CONCLUSION

Although the ear makes up a fraction of our total body surface area, the amount of different diseases seen is abundant. Because it is small and filled with many crevices, many patients neglect regular inspection of the ear. It is important as clinicians to be aware of examination, diagnosis, and treatment of this organ. Due to various conditions such as weathering, trauma, inflammation, immunologic and metabolic disorders, many challenging pathologic conditions of the ear exist. Clues to diagnosis may be found within the age of onset, clinical appearance, location, and symptoms of the lesion. Histopathology often helps to identify or confirm the diagnosis. Treatments are vast, but easily employed. Surgical techniques are often needed to confirm the diagnosis, ensure removal, and provide cosmetically pleasing results.

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