

## Skin and ear.

The ear and its covering skin have some peculiar characteristics and, moreover, some characteristics similar to those ones of the nose such as exposure, cartilaginous structure, less extensible skin and abundant sebaceous glands.

Some of these anatomic and physiological characteristics can influence several dermatological disorders. We should remember the abundant vessels and the susceptibility to vasomotor crises, the numerous sebaceous glands in the concha and the modified sweat glands in the bony portion of the external auditory duct, which are responsible for the secretion of cerumen and the presence of grooves and reliefs, which make the surface of the auricle very irregular. Finally, we should remember the retroauricular fold, which is often affected in many inflammatory constitutional skin diseases such as atopic dermatitis, psoriasis and seborrheic dermatitis.

The ear is usually uncovered and thus particularly exposed to the light and other physical agents. This is why it is affected by juvenile spring eruption. The ear is acrolocated and thus involved in some skin disorders with vasculitis component, such as erythema multiforme. Moreover, the ear is exposed and visible leading to all the consequences discussed with regard to the nose. The habit of piercing aimed at exposing earrings is a consequence of the visibility of the ear. Finally, the ear is exposed and seizable. This is why it is affected in a professional disease known as “ear of fighter”.

Due to the peculiar anatomy of the ear with its irregular surface (Fig. 1353), its tumors lie on different levels and their removal is thus more difficult. Other difficulties arise because of its rich vascularization. Also rebuilding operation is more difficult because of less extensible skin. The latter is also responsible for the severe pain

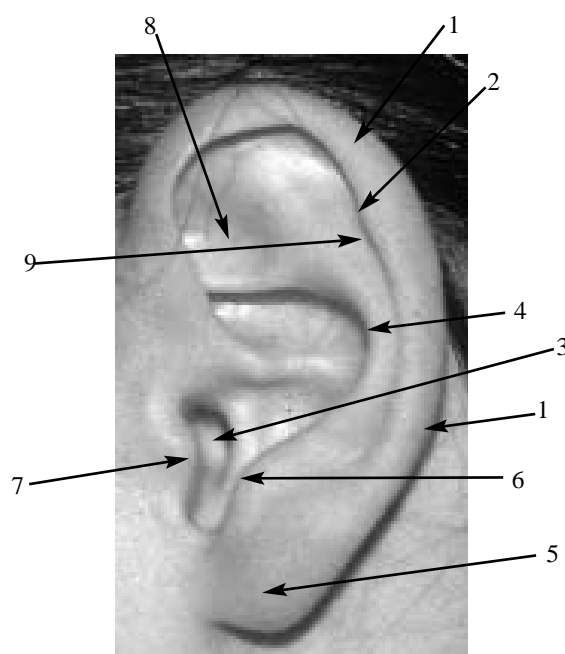


Fig. 1353: Left auricle observed from its lateral side. 1 helix; 2 tubercle of the auricle (or Darwin's tubercle); 3 concha of the auricle; 4 anthelix; 5 lobule of the auricle; 6 antitragus; 7 tragus; 8 triangular fossa; 9 scaphoidal fossa.

caused by folliculitis and foruncle affecting this site.

### Notes of embryology

These are useful to better understand the origin of numerous developmental anomalies such as branchial clefts and auricular sinuses and cysts located on this site and to more easily remember their distribution.

These lesions are developmental anomalies of the branchial arches. The gills are the respiratory organ in many fishes. The water and the oxygen

dissolved in the water enter from the mouth and go out from the gills. In the human embryo, as evolutive record, four paired pouches appear in the lateral pharyngeal wall in the fourth and fifth week. Simultaneously four grooves appear on the external surface of the embryo in direct apposition. In fish these two opposed structures meet and communicate to form gills.

On the other hand, in the human embryo only the first pouch and cleft remain in contact forming the tympanic membrane, whereas the other endodermal pouches and ectodermal clefts are separated by mesodermal tissue. However, on the lateral surface of the embryo, due to the contraposition of these structures, some ridges appear known as branchial arches. The latter, which consist of a cartilaginous nucleus surrounded by mesoderma, will give raise to the ear, lower jaw and neck structures. These ridges grow ventrally from both sides to join themselves on the mid line. The first branchial pouch gives raise to the Eustachian tube and the middle ear cavity, whereas the first branchial cleft gives raise to the external ear canal. The second branchial pouch gives raise to the palatine tonsil, but most of it is obliterated.

Causal factors of various type, acting between the fourth and the eighth week of embryonic life, interfere with the physiological process of obliteration of the branchial pouches and clefts, giving raise to developmental anomalies such as branchial clefts and auricular sinuses.

### Inherited disorders

An altered shape of the ear characterizes some inherited disorders such as neurofibromatosis, making, along with other dysmorphisms, the immediate diagnosis of neurofibromatosis easier (2, 5). The internal ear is particularly affected in central neurofibromatosis type II, whereas in peripheral neurofibromatosis type I plexiform neuromas can occur on the external ear (Fig. 1354).

Deafness or auditory alterations can occur in some multisystem inherited disorders which also affect the skin, as KID syndrome. In the latter auditory alterations are associated to keratitis

and ichthyosis (4). Hairy pinnae of the ear are linked to Y chromosome and therefore transmitted from the father to all his sons and only to them.

Branchial clefts, accessory tragi and preauricular sinuses and cysts are usually nevus malformations.

However, these anomalies can be transmitted as an inherited trait -Fig. 1355, 1356- (9).

### Nevi

Accessory tragus and preauricular cysts or sinuses are nevus malformations peculiar of this site. There are also melanocytic nevi, angiomas, nevi of the epidermis and epidermal appendages.

*Accessory tragus* is included among the developmental anomalies of the first branchial arch. The ventral growing of the first two branchial arches, which finally join themselves on the mid line anteriorly on the neck, explains the distribution of accessory tragus along a line starting from the tragus and arriving to the homolateral commissura of the lips or going down on the neck till the anterior margin of the sternocleidomastoid muscle.



Fig. 1354: Plexiform neuroma of the auricle in an 8-year-old girl with neurofibromatosis type 1.



Fig. 1355

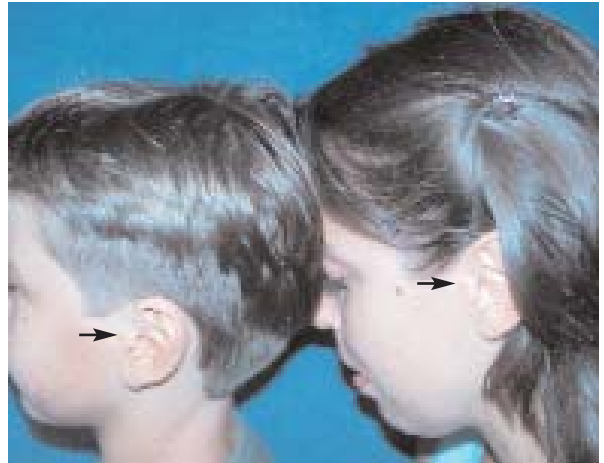


Fig. 1356

Fig. 1355, 1356, 1357: Accessory tragus may be inherited (Fig. 1355, 1356). More often it is sporadic, isolated or multiple (Fig. 1357), distributed according to a line starting from the tragus and arriving to the homolateral commissura of the lips or going down on the neck till the anterior margin of the sternocleidomastoid muscle.

Accessory tragus is a skin-colored nodule, sessile or pedunculated, isolated or multiple (Fig. 1357), mono- or bilateral and often covered by vellus hairs.

It is usually located in the preauricular region or less frequently in the mandibular or cervical region (Fig. 1358, 1359). At palpation accessory tragus can be soft or, more frequently, hard cartilaginous. Usually, accessory tragus is sporadic but familial cases are not exceptional (Fig. 1355, 1356).

The pathological findings of accessory tragus are characterized by a connective tissue structure



Fig. 1357

with abundant hairs often containing a central cartilaginous nucleus.

The surgical treatment is aimed at esthetical purposes and thus should be carried out only in the more visible cases. Attention must be paid in completely eliminating the cartilaginous nucleus, which is sometimes adherent to the surrounding tissues.

*Preauricular sinuses* and *cysts* are more rare developmental anomalies of the first branchial arch. Even more rare are the lesions located on the neck, usually in females and sometimes associated to deafness (6).



Fig. 1358: Accessory tragus of the neck.



Fig. 1359: Accessory tragus of the neck.

These anomalies are due to an incomplete obliteration of the branchial arch. They usually have a cutaneous mouth and a blind inner end. The sinus can be very long and from the anterior surface of the neck can reach the ear. These lesions are usually barely visible. This is why they are usually observed in case of infectious complications (Fig. 1360, 1361).

Histologically, the cavities are lined by a stratified squamous epithelium. Externally, the cavities are surrounded by connective tissue, that is particularly rich in skin appendages. The sinuses may arborize with cysts arranged around their tracts.

Clinically, when there is no infection, punctiform pits are barely visible, usually localized in front of the ascending limb of the helix. In case of infection, a secretion can be discharged from the mouth, which is surrounded by a more or less intense inflammation (Fig. 1360, 1361). A punctiform depression of the neck or the preauricular region may be a clue to the diagnosis of a deep inflammatory process of the external or middle ear.

When a fistula is complicated by infection, the surgical operation is mandatory, after the infection has been extinguished and the cavity has been probed in order to completely remove

the sinus tract. A computerized tomography is often useful (7).

Among the anomalies due to incomplete obliteration of the first branchial arch there are finally the *clefts* and *congenital vertical atrophic areas* of the mid line, which are sometimes associated to developmental anomalies of the tongue, lip, lower jaw and sternum.

Among the developmental anomalies due to incomplete obliteration of the second branchial arch there are the *lateral cervical cysts*, which are usually located under the mandibular angle. The latter arise late in the lower third of the neck, in front of the sternocleidomastoid muscle. The lateral cervical cysts can be blind ended or more or less deep till communicating with the pharynx. Infections can complicate their clinical course. Histologically, the lining epithelium can be squamous stratified or ciliated columnar or mixed.

Besides the isolated developmental defects, appearing as cysts or sinuses, the anomalies of the branchial arches can be part of complex syndromes, such as *Hallermann-Strieff syndrome* or *oculomandibular dyscephaly*, which is characterized by dwarfism, hypotrichosis, atrophic skin, beaked nose, etc., and *Goldenhar syndrome* or *oculoauricular vertebral dysplasia*.



Fig. 1360



Fig. 1361

Fig. 1360, 1361: Sinuses and cysts of the preauricular region complicated by suppurative infection.

With regard to the common nevus malformation, we have already mentioned the difficulties of surgical operation in this site. The latter regard all types of nevi.

Flat lateral angiomas of the face or port-wine stain affect the external ear when the second or

third trigeminal branch is involved (Fig. 1362). Due to the grooves and reliefs of the external ear also the treatment with dye laser at 585 nanometers wavelength (Fig. 1363), which is the first choice treatment for these lesions, is technically more difficult in this site.



Fig. 1362



Fig. 1363:

Fig. 1362, 1363: Capillary malformation -port-wine stain- partially affecting the third trigeminal branch with involvement of the auricle (Fig. 1362) and after 3 sessions of 585 nanometers dye laser (Fig. 1363).

Level with the lobule of the ear some sweat nevi such as eccrine spiroadenoma, which are characterized by thin papules in other sites, can exhibit a pseudotumoral development (Fig. 1364, 1365, 1366).

### Infections

Recurrent herpes simplex can affect the retroauricular region (Fig. 1367) or more often

the helix, sometimes bilaterally. Warts affecting the ear (Fig. 1368) are usually filiform in type. In this site the local intralesional anesthesia is more painful due to the scarcely extensible skin. Moreover, the rich vascularization of the auricle increases the difficulties of surgical removal.

Bacterial folliculitis and foruncle are frequent in the ear cavity, due to the high concentration of pilosebaceous follicles, and particularly painful due to the scarcely extensible skin in this site.



Fig. 1364



Fig. 1365



Fig. 1366

Fig. 1364, 1365, 1366: Sweat, eccrine spiroadenoma nevus of the temporal parietal region, ear (Fig. 1364, 1365) and right hemithorax (Fig. 1366). The lesions, which are micropapular on the thorax, get nodular on the ear.



Fig. 1367

Fig. 1367, 1368: Infections of various type can affect the ear. Among the viral infections there are cutaneous recurrent herpes simplex (Fig. 1367) and warts (Fig. 1368).



Fig. 1368

### Dermatitis

On the ear are localized both constitutional dermatitis, such as atopic dermatitis, seborrheic dermatitis and psoriasis, and allergic contact dermatitis. The differential diagnosis between

these constitutional dermatites is difficult in this site when based only on clinical features because scaling is less evident in the grooves of the ear and a slight exudation can be evident also in seborrheic dermatitis and psoriasis, which usually are not exudating. A typical sign is the



Fig. 1369

Fig. 1369, 1370: Under the lobule the differential diagnosis between atopic dermatitis and psoriasis is not always so easy as between the child in Fig. 1369 with atopic dermatitis and the girl in Fig. 1370 with psoriasis.



Fig. 1370

Fig. 1371, 1372, 1373: The difference between atopic dermatitis and psoriasis is usually more clear than in this girl. The latter, when aged less than 1 year, presented exudating atopic dermatitis of the ear (Fig. 1371) complicated by erysipelas-like dermatitis of the leg (Fig. 1372). Some years later the same girl presented lesions always more dry in the same site till the very evident diagnosis of psoriasis (Fig. 1373).



Fig. 1371



Fig. 1372



Fig. 1373



Fig. 1374



Fig. 1375

Fig. 1374, 1375: Allergic contact dermatitis to nickel of the sidepieces of the glasses (Fig. 1374, 1375) in an atopic young girl aged 12 years.

inflammation of the anterior superior margin of the lobule, level with the conjunction with the skin of the temporal region. We are dealing with a slight erythema, which can be the only sign of *atopic dermatitis*. In more severe cases the inflammation can break the epidermis (Fig. 1369) and cause linear crusted lesions and rhagadiform fissures. The latter are responsible for crying of the child when he/she is undressed and the lobule is forcedly pulled up. This sign, together with perioral erythema due to cheese and tomato and with persistent “cradle cap”, is included in the group of minor diagnostic criteria of atopic dermatitis and can be recalled even by history taking. Unfortunately, we are dealing with a generic sign of inflammation rather than a specific disorder. In fact, it is present even in psoriasis (Fig. 1370) and more rarely in seborrheic dermatitis. Therefore, the ear should not be considered, when facing the differential diagnosis between constitutional dermatites.

*Allergic contact dermatitis* due to nickel in children (Fig. 1374, 1375) and particularly in atopic children has been largely discussed with contrasting opinions regarding its prevalence. From a clinical point of view we could say that allergic contact dermatitis does not exist under

10 years, because we do not observe children aged less than 10 with this problem. However, this statement is simplistic because neither children nor adults are visited for this problem. In fact, diagnosis, treatment and prevention are usually carried out by the patient him/herself because of the immediate relationship between cause and effect. However, when questioning with patience in a systematic way a young population of teenagers with eczema, you can see that this problem usually starts around ten years and gets always more frequent with years. On the other hand, exceptionally parents report an itchy dermatitis of the lobule before seven years, in spite of the fact that piercing of the lobule is still extremely popular from the first years, still prevalently in the females.

The lobule of the auricle is not the only site of allergic contact dermatitis due to nickel. In fact the latter is even more frequent according to some Authors (8) level with the periumbilical region, especially in males, due to the contact with metal buttons. The wrist and the temporal parietal region are other frequently affected sites of allergic contact dermatitis to nickel. The latter site is affected due to the contact with the sidepieces of the glasses (Fig. 1374, 1375).



Fig. 1376: Erythema multiforme affecting the ear in a 4-year-old boy.



Fig. 1377: Juvenile spring eruption of the eyelid in a called up soldier (courtesy of dr. Vincenzo Petruzzellis).



Fig. 1378



Fig. 1379

Fig. 1378, 1379: Suppurative inflammation in a teenager. The latter was worried more about the possible closure of piercing than about the suppurative inflammation (courtesy of dr. Gaetano Scanni).



Fig. 1380

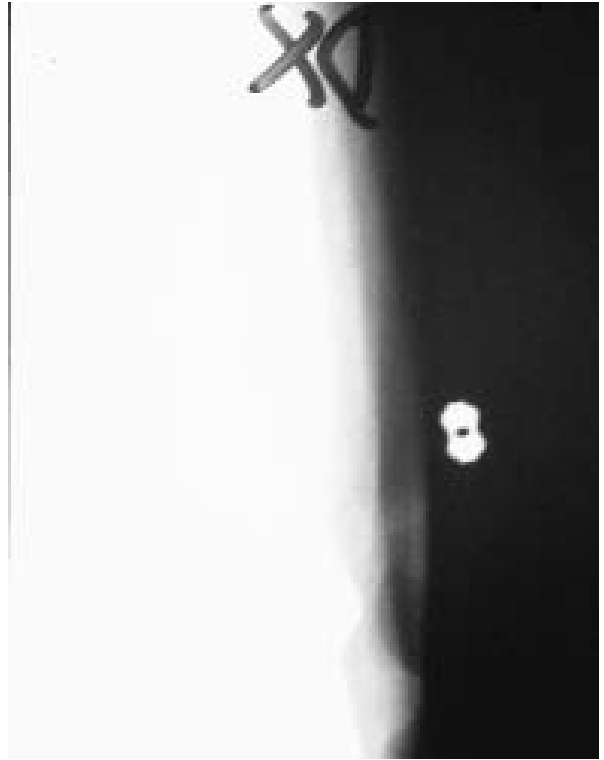


Fig. 1381

Fig. 1380, 1381: The girl as in Fig. 1380 was unaware that the lesion of her lobule was due to a foreign body, namely the stop screw of her earring, as X-ray film (Fig. 1381) showed before its removal.

A rare form of allergic contact dermatitis in teenagers is caused by plasters and glues, applied to the ear and the temporal parietal region aimed at temporarily remedying flap ears.

Besides constitutional dermatitis and allergic contact dermatitis, other two dermatites affect the ear. The first one is papular (Fig. 1376) and bullous *erythema multiforme*. In the latter disorder the ear is affected due to its acrolocated position, together with the face, hands, feet and knees.

The helix is characteristically affected in *juvenile spring eruption* (Fig. 1377). We are dealing with a photodermatitis, which mainly affects the helix, particularly its top region. It is characterized by pruritus and papular and vesicular lesions (1). The eruption characteristically affects in spring or summertime boys aged 3 to 12 years (3), although cases in conscripts have been reported. Juvenile spring eruption is a variant of polymorphic light eruption. Usually, it does not

affect females, whose ears are frequently covered by hair.

Complications of piercing, both chronic such as small cysts of the lobule and more often acute such as suppurative infection (Fig. 1378, 1379) affect the little girl and more frequently teenagers.

Another complication of piercing and earrings is the foreign body granulomatous reaction following the encapsulation within the skin of the lobule of the stop screw of the earring (Fig. 1380, 1381). In the latter case the affected patient is usually unaware of the responsible cause and the diagnosis is confirmed by an X-ray examination.

The “ear of fighter” (Fig. 1382, 1383, 1384) is another chronic granulomatous disorder affecting teenagers and young adults, who do all-in wrestling in an amateur or professional way. This disorder follows chronic traumas due to the competitive seizing of the ear by the opposing athlete.

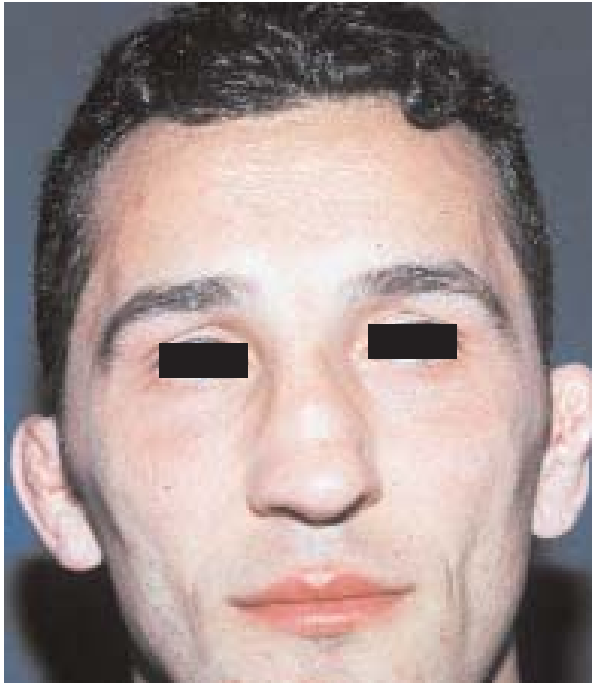


Fig. 1382



Fig. 1383



Fig. 1384

Fig. 1382, 1383, 1384: "Ear of fighter" in a 17-year-old teenager (Fig. 1382, 1383) and in his 18-year-old friend, who do all-in wrestling.

### Connective tissue disorders

*Discoid lupus erythematosus* is the most frequent connective tissue disorder affecting the ear (Fig. 1385). It is responsible for dyschromic and dystrophic scars with significant esthetical damage.

*Granuloma annulare* can also affect the ear. When the latter is the only site affected, the diagnosis is difficult, especially at the onset, when a hard papule is the only lesion. Later on, the latter enlarges peripherally with central resolution giving raise to an annular ring and making the diagnosis of granuloma annulare easier (Fig. 1386).

### Tumors

On the auricle (Fig. 1387) *hemangioma* does not grow significantly, maybe due to the compression exerted by the scarcely extensible skin. On the other hand, on the retroauricular fold (Fig. 1388) and on the preauricular region hemangioma often grows significantly and can be responsible for a marked esthetical damage, for instance pushing away the auricle from the

temporal parietal bone (Fig. 1388) when it is located in the retroauricular region, or pulling anteriorly the auricle in case of ulceration and consequent scarring retraction of a preauricular hemangioma (Fig. 1389, 1390).

These possible consequences should be taken into account during the evolutive phase of hemangioma and should suggest systemic corticosteroid treatment, aimed at preventing the devastating esthetical damage following ulceration and consequent scarring.

The intralesional corticosteroid treatment cannot be easily performed due to the pain and to the rapid passage of the blood in the vessels of hemangioma, which rapidly transfers in the general circulation whatever drug is injected in the lesion. We start systemic corticosteroid treatment with 1mg/kg prednisone and increase the dosage 0.5mg/kg every two days till obtaining the therapeutical response, consisting in the stop of the growth of hemangioma. This dosage should be continued throughout the entire growing phase of hemangioma, thus till the sixth-eighth month of life, possibly adjusting it to the growing weight and then should be tapered till withdrawing it within the ninth-twelfth month.



Fig. 1385: Discoid lupus erythematosus of the ear with dystrophic scar.



Fig. 1386: Granuloma annulare of the anthelix in a 14-year-old adolescent.



Fig. 1387



Fig. 1388

Fig. 1387, 1388: Hemangioma of the helix with multiple foci (Fig. 1387). In Fig. 1388 a hemangioma of the retroauricular fold in its growing phase pushed anteriorly and laterally the auricle.

*Pilomatricoma* is included in the group of benign tumors more frequently affecting the ear and even more frequently the preauricular region (Fig. 1391). Pilomatricoma derives from the hair matrix. Its clinical features varies according to the depth of the lesion, the presence of inflam-

mation and possible calcium deposits. Because of these possible events, pilomatricoma can be covered by flesh-colored, red-bluish (Fig. 1391) or yellowish skin. Characteristically, its consistence is hard stone and its surface markedly irregular.



Fig. 1389



Fig. 1390

Fig. 1389, 1390: Ulcerated hemangioma of the left ear and cheek in a 5-month-old baby girl (Fig. 1389). The regressing and scarring process following ulceration caused a clockwise rotation of the auricle and an anterior traction of the lobule.



Fig. 1391: Pilomatricoma of the ascending limb of the helix in a 5-year-old girl.

Under and behind the lobule there are epidermal cysts of sebocystomatosis in the teenagers and young adults. This disorder responds to retinoids.

Among the malignant tumors we remember basal cell and squamous cell carcinoma, much



Fig. 1392: Juvenile xanthogranuloma of the auricular lobule in a 6-month-old boy.

more rarely melanoma, in children with xeroderma pigmentosum (Fig. 1393).

Finally, we mention the purpuric micropapules of Langerhans cell histiocytosis, often localized in the retroauricular region (Fig. 1394) and thus simulating seborrheic dermatitis.



Fig. 1393: Poikiloderma and dyskeratosis (arrow) of the ear in a 12-year-old girl with xeroderma pigmentosum.

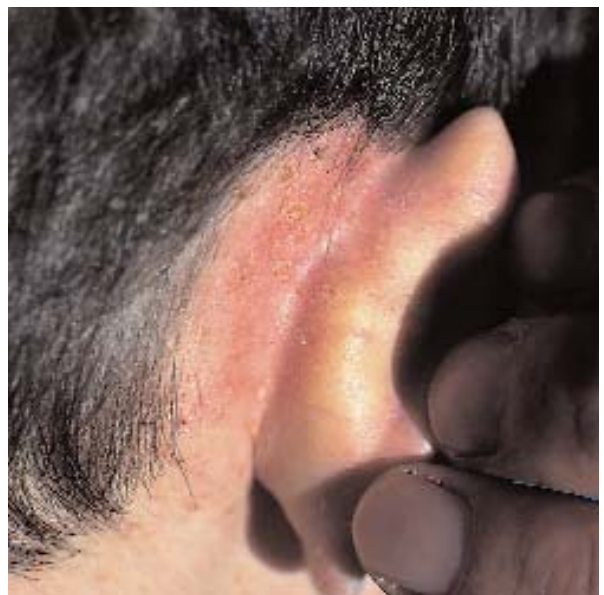


Fig. 1394: Purpuric and crusted isolated papules of Langerhans cell histiocytosis type Letterer-Siwe.

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