

following Hebra's doctrine, class as more or less accidental concomitants of other processes; for instance, the forms of ulcers due to large excoriations (especially from pediculi vestimentorum).

2. Especially syphilitic gumma in all its different stages of development which pretty closely correspond to those of a scrofulous infiltration. Essential to the differentiation are here:

*a.* The absence or presence of former symptoms of syphilis. If syphilis be absent in the history or in the examination, this furnishes a diagnostically valuable support. But inversely, if there was a former syphilis, the possibility of the coincidence of syphilis and tuberculosis is still to be taken into consideration.

*b.* Scrofulous abscesses are almost exclusively found as concomitant phenomena of general scrofulous disease, while gummata frequently appear in otherwise healthy persons (syphilis in a late stage).

*c.* The scrofulous neoplasms are generally more numerous than gummata. The latter, moreover, show a preponderating tendency to localize on the forehead and the portions of the skin covering the long bones.

*d.* The scrofulous abscesses always spring from the subcutaneous tissue, very frequently from the lymphatic glands. Gummous infiltration is as often cutaneous as subcutaneous, and gummata have nothing whatever to do with the glands.

*e.* The different appearance of the formation also decides; in the initial stages, gumma is harder and firmer than the scrofulous mass of granulation. In the stage of softening, fluctuation is slighter in gumma. In the ulcerative stage, the differences are still more obvious; here a hard, firmly infiltrated gumma mass, which does not collapse even after opening, with but very slight gummous secretion; there an abscess with relaxed borders, loose base, and equally flabby environs, which at once collapses with the discharge of the thin, fluid pus.

*f.* The ulcerous forms of syphilis are clear cut, usually painful ulcers, with steep, firmly infiltrated borders; scrofuloma ulcerosum, however, presents itself as a painless ulcer, with thin, soft, irregular, undermined borders, which, besides, enlarges much more slowly than the serpiginous luetic ulcers.

*g.* Finally, the exhibition of potassium iodide may settle the question. This agent causes rapid absorption of the gummous neoplasms, while it scarcely influences the course of the scrofulous new-formation.

*Treatment.*—This consists essentially in a roborant general treatment directed against scrofulosis: cod-liver oil, syrup of iodide of iron, etc. The local treatment is chiefly surgical: removal of the caseated and softening infiltrations with the sharp spoon; ablation of the thinned borders with knife and scissors, and a stimulating dressing to the granulation of the bottom of the ulcer. Wine of camphor, silver nitrate ointment (one to two per cent), best iodoform dusted on in powder, or dissolved in ether, or as an ointment dressing (one in fifteen), etc. At any rate, as long as there is profuse suppuration, sufficient egress must be given to the pus. Atomization of iodoform-ether (1:15), or camphorated iodoform ether, is especially appropriate for sinuous ulcerations which are accessible with difficulty (in the nasal or oral cavity). Lymphatic glands must eventually be removed, osseous fistulæ scraped out.

In the case of these scrofulous abscesses and fistulæ, we must bear in mind that the granulating surfaces are often covered with granulation epithelium (Friedländer), which of course prevents the direct union of opposite wound surfaces, and accordingly must be removed (with the sharp spoon).

### 3. *Lupus.*

The morbid picture of lupus belongs to the most manifold and changeable of all forms encountered in dermatology.

The morbid process consists in the fact that (either by direct infection from without or from within from subcutaneous foci) the skin is penetrated by a specific virus at present still unknown, but, we suspect, to be identified with the bacilli of tuberculosis. This virus acts in the first place by exciting inflammation, and thus gives rise to the formation of small inflammatory patches situated along the vessel walls. These small neoplasms, composed of inflammatory granulation tissue, appear on section as little spherical nodules (granuloma, lupus nodules), and are deposited in midst of a connective tissue likewise diffusely infiltrated with inflammatory cells. The virus subsequently exerts its specific qualities in two directions.

1. It influences the course of every single new-formation produced by it.
2. It causes peripheral migration of the neoplasms by spreading also into the healthy periphery.

Ad 1. In the centre of every nodule (*i. e.*, a spherical accumulation of small lymphoid cells which are embedded in a fine fibrillary connective-tissue mesh-work, and between which project sparse blood-vessels from the periphery) the virus produces destruction of the cells as well as of the fibrillary basement tissue; a specific necrosis develops, a "caseation," while toward the periphery of the nodule progressive developmental phases are found in the cells: the formation of large epithelioid and giant cells. But even these are only temporary formations, because they, like the central cells, gradually succumb to the necrobiosis extending outward.

Ad 2. While these processes take place in the single lupus nodule which has been called very happily the "primary efflorescence" of lupus, the virus has migrated into the neighborhood, and there has again produced "nodules," on which the old process is repeated. These, too, caseate, and, as the nodular masses all perish one after another, not only the first-attacked portion of tissue is destroyed, but likewise its surroundings which coalesce with the new-formation both in width and in depth, and perish.

With this most important point—formation and disintegration of the lupus nodules under the influence of a virus spreading into the periphery—other factors come into consideration, which, though in themselves of secondary importance, very materially modify the clinical picture of lupus. These are:

1. The manner of the spread of the lupus poison and the lupus nodules (primary efflorescences) produced by it.
2. The participation of the internodular connective tissue which is either limited to a simple, temporary infiltration with inflammatory cells, or may lead to node-like hyperplasia, at times circumscribed, at others general, approaching elephantiasis.
3. A special activity of the papillæ, by which eventually papillary, horny excrescences are produced.

As regards the epithelium, however, it participates to a slight extent, and contributes little to the specific character of the morbid picture.

Therefore, by lupus vulgaris we understand a chronic cutaneous disease which arises from the introduction of small patches of infiltration into the tissue of the skin, and progresses by the peripheral spread of ever renewed similar patches, while the older ones perish by slow absorption, or by necrobiosis leading to ulceration. The variable participation of the connective tissue and the papillary body produces different forms, on the description of which we shall now enter.

*Lupus maculosus*.—The primary efflorescences, the lupus nodules, are perceptible as small spots the size of a millet-seed through the epidermis, which is slightly depressed at these places situated in the midst of a cutaneous surface which otherwise appears quite normal. The color of these spots is a yellowish-brown, the intensity of which can be only imperceptibly lightened by pressure with the finger. The skin above the spot is smooth or very slightly scaly; it is glossy, especially on side illumination, because the normal linear tracing is absent. The most characteristic feature is the consistence. A light pressure with the blunt knob of a sound suffices to penetrate through the epidermis-layer into the cell-mass and to bury the knob in the depth; while otherwise the epidermis is impenetrable to a blunt instrument even on strong pressure.

The lupus nodules are always situated in the connective-tissue part of the skin, both in healthy, formerly intact portions, and in old cicatrices in which the color contrast of the yellowish-brown infiltration with the whitish scar, of course, is especially evident.

*Lupus maculosus* is always the first stage of the development; it is found, therefore, either as the first sign of the affection generally, or in the periphery of older foci, or as a relapse in cicatricial surfaces, usually in small, more or less circularly arranged groups.

No subjective inconveniences are connected with the development of the lupus nod-

*Lupus exfoliatus*.—In the further course, which is very chronic, there are now developed very numerous, closely packed nodules, the final result being a generally circular lupous surface, two to three centimetres in diameter, brownish-red to brownish-yellow in color. The central oldest portion is usually already in a state of regressive metamorphosis (fatty degeneration, caseation, and cicatrization), hence darker and depressed, while toward the periphery the more recent lupus efflorescences are lighter in color, and often united into a slightly elevated, wall-like crown. Besides this closely packed conglomeration of lupus nodules (which, therefore, do not represent a uniform diffuse infiltration), the healthy surroundings are sprinkled with isolated, most recent nodules.

The deep-seated infiltration, having led to the destruction of the papillary body and flattening of the cones of the rete, robs the surface of such extensive patches of lupus of its linear tracing; the surface, therefore, is glossy. Withal, the adherence of the cornified epidermis cells is impaired, hence the surface is scaly, rough, and fissured. The transverse diameter of the skin is thinned, and consequently becomes easily wrinkled and folded. This quality of superficial desquamation of the epidermis has given this developmental form the name of *lupus exfoliatus*.

Of late I had two opportunities of seeing lupus forms belonging under this head, which were spread over large surfaces. In the course of many years (twelve to eighteen), the affection had involved almost the entire thigh and the whole of the gluteal region, without having led to ulceration or healing at any spot. The gradually enlarging affection represented a uniformly glossy, slightly scaly, brown surface, covered with wrinkled epidermis; at first sight, it looked even more like an abnormally colored psoriatic surface than like lupus. Some primary efflorescences, however, soon determined the diagnosis, which was confirmed by the microscope. The latter demonstrated a very copious infiltration with groups of granulation cells (with very large and numerous giant-cells) in the uppermost layers of the corium, and in the papillæ between the cones of the rete. In the subcutaneous tissue the lupus nodules were very sparse.

*Lupus exulcerans*.—Now and then healing processes take place, *i. e.*, melting down and subsequent cicatrization without destruction of the epidermis, a kind of subcutaneous scar-formation (cicatricial atrophy). Usually, however, the intensity and acuteness of the necrobiotic processes in the lupous tissue, the caseation and softening, are so great,

and by irritation from without more vivid inflammatory phenomena are superadded to such a degree that the normal epithelial covering is destroyed, and the disintegrating lupous infiltration is freely exposed. A more copious secretion, "suppuration," of this wound-surface at once ensues, so that now we speak of it as an "ulcer": *lupus exulcerans*.

What is the nature of such a lupous ulcer? The base is usually covered with yellow and (by admixture of blood) brown, not very thick, flat crusts. If these be lifted and the underlying puriform fluid, consisting of pus-cells and detritus, removed, we expose the red, somewhat irregular base covered with flat granulations; it bleeds easily and shows the same consistence, at once yielding to every instrument, as the single lupus efflorescences. The borders imperceptibly blend with the base of the ulcer; they often exhibit, for a distance of several centimetres into the surroundings, the brownish-yellow color and the soft consistence of lupus skin, at times they are also slightly elevated, wall-like (by more massive infiltration).

*Lupus serpiginosus*.—The granulations, that is to say, the freely exposed corium and subcutaneous connective tissue infiltrated by lupus, now gradually disintegrate, and are replaced by newly forming, persisting connective tissue, which is gradually covered with epithelium from the neighborhood; thus a further stage is reached. The lupus heals, and very dense, firm, irregularly distorted, retracting cicatrices result. Such healing, however, is effected only in the centre, or at one segment of the circular ulcerating surface, namely, where the lupus poison has died, and, therefore, no longer prevents the tendency to recovery. But it is all the more active in the still affected peripheral zone; there it "creeps" onward (*lupus serpiginosus*).

But under some circumstances the inflammatory neoplasia of the connective tissue in the base of the ulcer may preponderate over the necrosis; greatly elevated proliferations of granulation then form, which not rarely may organize into some permanence, become covered with skin, and thus remain unchanged for years. In other cases the proliferations are more relaxed, more friable, and bleed easily. Under the microscope we distinguish a tissue built up of young connective tissue, more or less interspersed with lupus nodules. This connective-tissue formation, proliferated under the chronic irritation of the lupus poison, is termed *Frambœsia luposa* or *Lupus frambœsioides*.

The same process, however, is found not only springing from the base of the lupus ulcerations, but, more frequently, as a developmental form of *lupus maculosus* (still covered with epidermis). In this case are formed chiefly firm neoplasms composed of a connective tissue representing different degrees of development of the inflammatory connective-tissue new-formation and almost identical in structure with ordinary elephantiasis. We then observe brown or brownish-red, soft or firmer elevations from a pea to a nut in size, either spherical or like low flower-beds; their surface is smooth, or slightly scaly, often excoriated and covered with thin crusts. If the development of the papillæ—*i. e.*, the growth of its ascending vascular loops with corresponding depression of the cones of the rete—is especially prominent, verrucose nodes arise which permit the recognition of the papillary structure on the surface: *lupus papillaris, verrucosus*.

But all these neoplasms, despite their frequent persistence for years, are not permanent. Sooner or later, with the interspersed lupus nodules, the connective tissue inclosing them likewise perishes.

We have thus far become acquainted only with extension of lupus by contiguity. Besides, we speak of a *lupus disseminatus (discretus)* when the several foci appear on

different parts of the body either simultaneously or in succession, in which case every patch may independently change into a lupus serpiginosus.

As to the extension into the *depth*, the virus migrates in the lymph channels, around the ducts of sudoriparous glands which penetrate deeply, etc., progresses along the vessels, penetrates the connective tissue, stopping as a rule at the firm fasciæ and the bones. Now and then, however, the periosteum is also destroyed, giving rise to more or less superficial necroses of the bone. Cartilaginous tissue is very readily attacked by lupus and penetrated by the neoplasm.

In by far the majority of cases of lupus, the disease shows itself first on the external skin, and thence extends to the mucous membranes (mouth, pharynx, larynx, conjunctiva, etc.). In rare cases primary lupus of the mucous membranes has been observed, and then, as a rule, the small brownish-red "nodules" are not to be seen. Small whitish exfoliations of the epithelium show themselves in the livid-red, slightly thickened mucosa. Subsequently they change into small warty prominences which either still exhibit the whitish, thickened epithelial coating or have a red, easily bleeding surface.

The lupous process may occur on any part of the body, but attacks the face and the extremities with the greatest frequency. Usually the patches are isolated, not exceeding the size of the palm of the hand; only in rare cases it covers the entire face, neck, and parts of the trunk.

*Lupus of the Face.*—It begins either in isolated flat macular form on one or both cheeks, and usually remains for a long time unnoticed by the patient; or else, on the nose in the form of lupus tumidus, tuberculosus, gradually leading to swelling and livid discoloration. While the central portions heal with the formation of cicatrices, the process spreads in the neighborhood either serpiginosus or by peripheral extension of fresh isolated groups of nodules, involving the upper lip, the forehead, and eyelids. Then follows a gradual confluence of older, originally isolated patches, thus leading to extreme disfigurement. The cicatrized surfaces are not only themselves irregularly distorted, but they also cause ectropion of the eyelids and lips which is rendered all the more conspicuous by the swelling, partly œdematous, partly by lupous infiltration, of the surroundings. The disfigurement becomes still greater by the interspersed ulcerations covered with crusts, and the variable color of the several patches, changing with the age of the process. After the infiltration has spread in the deeper subcutaneous and intermuscular layers of tissue, a more regular swelling of the livid discolored surface results, often without external cutaneous manifestation of the deep-seated infiltration which becomes visible at the surface only in the course of time, by means of long fistulous tracks with irregular ulcerous openings. The morbid picture is completed by the prominence of the groups of glands on both sides of the face and in the submaxillary region, the softening and caseation of which sooner or later implicates the skin and cause fistulæ and indolent ulcerations (scrofuloderma). It is especially noteworthy that the cutaneous affection over these "scrofulous" abscess cavities bears the typical character of lupous disease. Lupus of the forehead usually presents a flat macular form, without any particular tendency to disintegration. But often it extends to the scalp (where it may lead to caries of the cranial bones), also to the upper eyelids which become ectropic.

But the most serious deformity is caused by lupus of the nose and upper lip.

Lupus of the nose may be a *L. maculosus* and *exfoliatus* which has deposited a flat infiltration on the ala or the dorsum. With the absorption of the neoplasm the basement tissue also perishes, and as a result there is shortening, shrinkage, mutilation,

diminution of the nose. When the affection has been pretty uniformly distributed over the surface, the organ is reduced in all its diameters, its point is retracted as by a bridle.

Or else the volume of the nose is increased if the form be of the tubercular variety, with more copious neoplasia of more acute onset. The nose now is brownish-red, irregularly swollen with knobby elevations, the surface is rough, usually with shallow ulcers, and covered with thick crusts and dirty scabs. In the neighborhood, isolated lupus nodules are seen scattered over the healthy tissue. The pituitary membrane also becomes implicated. The membranous and the cartilaginous septum, also the cartilage of the roof of the nose and the alæ, become infiltrated, thickened, and covered with inspissated masses of secretion which not rarely completely occlude the nostrils. But when the disintegration begins spontaneously or when the physician removes the lupous masses mechanically, we get an insight into the extent of the destruction. Instead of the club-shaped enlarged nose, not rarely a small remnant, confined to the osseous part, remains behind; the membranous and cartilaginous portions are to a great extent missing. Even now, however, the process does not come to a standstill, but migrates backward on the mucous membrane of the nasal cavities and the septum, leading to the formation of enormous crusts on the ulcerated surfaces, even to the sequestration of the bony parts of the nasal framework, of course with corresponding diminution of the air passages.

The destruction of the membranous and cartilaginous septum narium comes under observation also with intact skin, so that the nose is depressed *in toto*, the roof being straight.

Lupus of the *upper lip* very soon extends to the mucous membrane, where it causes an uneven surface resembling granulations. The lip is enormously swollen and everted, irregularly traversed by rhagades, fissured, bleeding easily. Should such a rhagade-like ulcer heal, the deep scar and band-like retraction increase the disfigurement. Finally the mouth can no longer be closed, thus still further favoring the ulceration of the mucous membrane. After the process has extended for years, those terrible deformities result which nowadays fortunately come under observation only from regions far removed from medical aid.

Lupus also extends farther on to the mucous membrane of the oral cavity. The gums, the mucosa of the soft and hard palate, become soft, tumefied, the investing epithelial layer loosened and whitish, until bleeding defects result which are sometimes covered with proliferating granulations of great persistence, sometimes soon give place to a superficial cicatrix. But when the lupus penetrates more deeply, very extensive ulcerations result which finally terminate in the permanent destruction of the velum palati, the uvula, the tonsils, etc. The epiglottis, too, as well as the mucous membrane of the larynx, become involved, then showing a surface with slight tuberosities on a diffusely swollen and greatly reddened base. Ulcerations succeed and the formation of contracting cicatrices terminates the process which leads to extreme hoarseness, attacks of dyspnoea, and finally total aphonia.

*Lupus of the auricle*; great swelling of the whole of the pinna very soon develops. Specially characteristic is the enlargement which the lobule undergoes, making it depend from the cartilage as a soft, pear-shaped tumor; the epidermis may remain intact for a very long time, and gives place to a uniform, superficial ulceration at a late period. The auricle and the neighboring integument may be almost totally destroyed, the subsequent cicatricial formation occluding the external auditory meatus.

*Lupus on the neck* develops either in direct continuity from the face, in the shape

of shallow ulceration, and final cicatricial formation drawing the head forward; or else isolated patches developing over softening lymphatic glands in connection with so-called scrofulous ulcers (see above).

*Lupus of the trunk and the extremities* usually occurs in a typical serpiginous form, and, in the course of years, covers large portions of the body. Of greater interest are the patches of lupus situated on the *joints*, which may give rise to disturbances of motility by the formation of ulcers and subsequent cicatrization. Flexion and extension are then often very materially impaired. On the hands, the bones are likewise implicated; caries and necrosis, deep ulcers and firm cicatrices arise, the fingers become curved, shortened, phalanges are sequestered and cast off, subluxations form, etc. Often enough we are compelled to perform amputation. Otherwise, lupus of the extremities is distinguished rather by the combination with a hyperplastic involvement of tissue; we find massive tumefactions, usually covered with crusts, diffuse and circumscribed thickenings (dorsum of the hand); a diffuse hypertrophy of large segments (feet, legs), comparable with elephantiasis, develops; moreover, the form of lupus papillaris s. verrucosus is nowhere as frequent as on the extensor surfaces of the elbow and knee joints, on the dorsum of the foot, and around the lupous ulcers on the legs. "The limb is thickened so as to resemble a wooden leg; the skin with the subcutaneous connective tissue, the soft parts, and bones is changed to a rigid mass, not firm, irregularly nodular on its surface, here and there glossy, tense; at other points covered with thick, dirty epidermic callosities, on still other parts, set with warty excrescences and spinous outgrowths."

*Lupus of the Genitals.*—On the male genitals we usually find infiltration of the prepuce to which the lupus has migrated from the neighborhood (buttocks, thigh). On the female genitals, the lupous ulcerations are of special importance, on account of the differential diagnosis from other forms of ulcers. Their occurrence is comparatively rare.

*Lupus of the Mucous Membranes.*—This form has nearly always migrated to the mucosa from the contiguous skin. "Lupus nodules," in as distinct an isolation as on the skin, are, of course, not recognizable; there are rather papillary excrescences with whitish epithelial opacities above the small spherical lupus infiltrations which are not recognizable microscopically. The excrescences unite into patches with roundish serpiginous borders, finally breaking down into ulcers. These ulcers are distinguished by their persistence, because the cicatrization is retarded by relapses in the granulations. Thus there are frequently immense proliferating granulations (frambœsia luosa) which disintegrate late and leave unsightly tumid cicatrices. Primary autochthonous lupus of the oral mucous membrane, of the larynx, and of the conjunctiva is exceedingly rare. Still, a large proportion of these cases is certainly overlooked. Many so-called eczemas of the Schneiderian membrane are probably nothing else but lupus, without our being able at the time to verify such a supposition unless more extensive destruction of the septum or the appearance of lupus on the integument furnish further diagnostic landmarks.

A very frequent complication of lupus is erysipelas, in consequence of the ulcerating surfaces favoring the entry of erysipelas-cocci. Some authors claim to have observed rapid healing of lupus by the erysipelatos process.

**COURSE.**—Most cases of lupus develop in childhood, between the third and tenth years; they begin with isolated, small patches, which, as a rule, escape notice. In this way, the small lupous infiltrations on the extremities remain for years, or at most give rise to superficial ulceration and cicatrization, until about the time of puberty, when

suddenly a rapid development occurs with destruction of the affected portions. After puberty the occurrence of lupus is rare.

In reference to the number of places of eruption, lupus appears either in one single patch, or simultaneously in a number of disseminated foci. In the latter case, of course, the prognosis is more unfavorable.

The spread of the process is always very slow and extends over many years before a more important development in extent or depth ensues. Lupus maculosus and exfoliatus are much more insidious and hence more benign than the far more rapidly extending lupus exulcerans in which suppuration is associated with the destruction of the tissues.

Owing to the exceedingly chronic course in most cases, the influence of the disease on the general health is little or not at all injurious. Only the complications resulting from the suppuration of the glands, the formation of long fistulous ulcers, the erysipelata and lymphangitides, etc., starting from lupus ulcers, may in a few—certainly rare—cases give rise to danger.

As regards the **PROGNOSIS** of the local process, local healing, both spontaneously and after therapeutic measures, takes place; but in every case there is danger of relapse. Very often we see in the midst of large cicatrices (old lupus ulcers) the primary efflorescences spring up anew; even in large transplanted flaps from the healthy forehead the development of lupus nodules, by immigration from the nose, is of frequent occurrence.

Lupus is far more frequent in females than in males.

**ETIOLOGY.**—According to our hypothesis, lupus is nothing but tuberculosis (s. scrofulosis) of the skin, produced by the bacillus of tuberculosis. Only the localization of the bacillus in the skin, the relatively rare involvement of other organs, give rise to the peculiarity of lupus as opposed to the other forms of tuberculosis.

Qualitatively (probably) the bacilli are the same, only quantitatively there is a difference which is intensified by the less favorable nutritive conditions in the cooler skin. Complications with tubercular affections of other organs—glands, joints, bones, even with analogous cutaneous diseases, scrofuloderma ulcerosum, etc. (scrofulosis)—are very frequent; their absence has no argumentative force against the tubercular etiology.

Lupus has nothing in common with syphilis in any direction. Both diseases may run side by side in the same individual.

**DIAGNOSIS.**—In the diagnosis of lupus the following main points have to be adhered to:

*A. From the History:*

1. The beginning in early childhood, or before puberty.
2. The very slow, insidious course.

*B. From the Objective Condition:*

1. A new-formation consisting of soft, friable granulation tissue, situated *in* the skin or rising but little above the level of the external surface.
2. The formation of "primary efflorescences" in the neighborhood of older foci; these small, yellowish-brown, easily depressed nodules scattered in the tissue are the most valuable diagnostic sign of lupus.
3. The tardiness of the development from the single lupus nodule to the broader conglomeration of nodules which again requires months to break down into an ulcer.