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BLISTER DERMATOSES

There are a group of diseases of different genesis but joined according to the monomorfiness eruptions (blisters), rarely combined with other morphological elements (During"s dermatosis)

- This group includes acantholytic pemphiqus, non-acantholytic pemphigus, congenital bullous epidermolyses, During's dermatoses.

Acantholytic pemphigus (real)

This is a disease with a chronic wavy course with development of blisters on the unchanged skin and mucous membranes tending to generalize with the aggravation of the patient's condition. It oftener develops at the age of 40-60, 70% of patients are women.

- In the development of acantholytic pemphigus genetic factor is of great importance.
- However, triggered role in case of predisposition is played by food factors (mustard, nut ceshu, mango and some other diets).

Etiology

of acantholytic pemphigus is still unclear.

- In case of pemphigus some metabolic disturbances (water-salt exchange, oxidizing-reducing enzymes, exchange of thyrosine and so on) have been found out, as well as endocrine pathology (dysfunction of sex glands, adrenal glands).

- But many authors consider that the filterable virus or a group of biologically close viruses (so-called "slow viruses") are etiological factors, and immunological, endocrine, metabolic and enzymes disturbances play only pathogenetic role.

- At present the leading role in patho-genesis is played by autoimmune changes.
- Autoantibodies (Ig of G class), deposited in the region of tonofilaments , connecting spinous cells are produced by the patient's cells.

- As a result formation of numerous prolonged cracks in the body of epidermis, filled with serous exudates (the process is called "acantholysis" after which the disease has been called) with formation of intraepidermal blisters takes place as a result.

Clinic.

Depending upon predominating clinical manifestations 4 forms of a real acantholytic pemphigus are distinguished:

1. Vulgar pemphigus
2. Vegetating
3. Leaf like (exfoliative)
4. Seborrhea (Senir-Asher syndrome).

Vulgar pemphigus

It seen in 75% of all cases of acantholytic pemphigus and are marked by a malignancy of it's course.

Before the use of corticosteroids in the past the patient's death occurred in the period of some month up to 2 years.

The clinical manifestations

of the disease are blisters appearing on the unchanged or slightly hyperemic skin.

- The size of the blisters varies from a pea-size to the Greek nut.
- Dermatosis begins, as a rule, in the oral cavity and then eruptions appear on the skin of the trunk, folds, genital organs, face.

- The contents of the blisters are serous at first, in 1-2 days it becomes "lemon", rarely hemorrhagic and cloudy. First, the coverings of the blisters are tense, dense and after that they become flabby and change their form to the pear under the weight of exudate.

- On pressure of the covering of the blister one can see the liquid to exfoliate the close laying healthy areas of epidermis and the area of the blister increases in diameter (the symptom of Asbo-Hansen).
- Acantholysis (the loss of connection between the spinous cells of epidermis) is at the base of one more clinical symptom of Nickolsky

- If we pull the scrap of the blister covering the wedge-like exfoliation of a visibly healthy epidermis will occur (something it has the length of dozens of centimeters) -the edge symptom of Nickolsky.
- On fractioning of the visibly healthy skin or at on it's scraping with a blunt instrument the "shifting" of the upper layers of epidermis (a remote symptom of Nickolsky) occurs between the blisters or erosions.

- On rupture the blister covering bright moist erosions occur. The characteristic feature of erosion is it's slow healing.

- Due to the secondary infection development the surface is often covered with purulent crusts, which are exfoliated after epithelization

- The most patients do not have any subjective feelings during appearing the blisters but numerous erosions cause burning, pain on swallowing and movement.
- The symptoms of acantholysis (Asbo-Hansen, Nickolsky) are positive only in the phase of exacerbation.

- There are several phases in the course of pemphigues:
- ***The initial stage.***
- ***The phase of exacerbation***
- ***Terminal stage.***

Vegetating pemphygus

is a variety of a real pemphygus and occurs rarer.

- The process begins from the mouth cavity.
- On the skin eruptions are oftener localized in the area of the axillaries pits, inguinal folds, perineum.

The blisters are less in dimension than in case of a vulgar pemphygus.

- After the opening of the blisters due to the irritation and maceration of the skin vegetations, the expansion of the papillary layer of derma and epidermis on 1-2 cm above the skin level develops in the folds.

- On the surface of the red vegetations the abundant discharge accumulates which quickly decays producing an extremely unpleasant smell.
- Without an adequate therapy the disease ends lethally in some weeks or months.

The leaf-like pemphygus (exfoliative).

In this form the more or less expressed blisters are seen at the beginning.

- Later, at the increase of symptoms of acantholysis a universal skin defeat develops which resembles exfoliative (leaf-like) erythrodermia.

- In this, vast areas of the skin look like continuous erosions or crusty stratifications resembling flaky dough.
- The course is severe. In children prognosis is worse.

Seborrhea pemphygus (Senir – Asher syndrome).

The clinic of seborrhea pemphygus reminds the symptoms of lupus erythematodes, vulgar pemphygus and seborrhea dermatitis.

- The initial symptoms appear more frequently on the skin of the face, head forming erythematous foci covered with scaly crusts resembling "butterfly" of the lupus erythematodes.
- The foci of the lesions of the face may exist from several months till 5-7 years and only later generalization of blisters eruptions on the trunk and extremities.

Diagnostics

is based on a typical clinical picture, symptoms of acantholysis (Nickolsky and Asbo-Hansen) and on the obligatory revealance of acantholytic cells of Tzank in the smears imprinting from the bottom of the erosions or from the lower surface of the covering of the blisters.

- Acantholytic cells are degenerative changed the spinous cells of epidermis.
- They are smaller than the normal cells, nuclei are big, dark-colored, occupy almost the whole cell with a thin band of light-blue cytoplasm.

- On of the diagnostic methods is to reveal by the immunofluorescent method the deposits of lumiscenting IgG in the region of intercellular contacts of spinous epidermocytes.

Treatment

- According to the life evidences systemic corticosteroids are administered.
- The treatment begins with the maximal doses (tabulated prednisolone from 70 to 120 mg per day, in injections-from 90 to 400 mg)
- The cessation of eruptions, a decrease of exudative symptoms is the sign to decrease the daily dose of corticosteroids.

- The principle of the further treatment consists of determination of the daily minimal dose of steroids in which new eruptions do not appear (usually it is 1 - 5 tablets of prednisolone per day).

- This is so called supporting dose which is given to the patients during a long period of time, often during the whole life.
- Periodically on the base of the supporting dose administration the patients develop exacerbation, the way of treatment in this case is the same.

- Regarding the water-electrolyte impairment of metabolism and also to avoid side-effects of long-term administered anabolic steroids are recommended -normalization of the proteum exchange (nerabol, retabolyl, and others), potassium preparations (due to its quick excretion from the body due to steroids intake), calcium (due to avoid to develop osteoporosis).

- Immunostimulators and antibiotics (the expressed immunosuppressive action of steroids), vitamins (C – the impairment of the vascular walls integrity, B – normalization of metabolism), in case of necessity – infusion therapy, haemosorb-tion, plasmopheresis, haemotransfusions.

Local therapy.

- Baths of manganese, painting the erosions with water solutions of aniline paintings, rinsing the mouth cavity with furacillin, steroid and epythelizing ointments – vulnuzan, solcoseryl, panthenol.

During Herpetiform Dermatosis

- This disease is considered as chronically recurrent allergy-toxic dermatosis, revealed by polymorphism of its eruptions on the skin and the mucous membranes.

- According to the modern theory gluten enteropathy of the jejunum (the other name is gluten disease) is at the base of its pathogenesis.
- The impairment of absorption is due to fermentation causing sensibilization to gluten – one of the constituents of cereal plants protein.

- Thus, During"s dermatosis it is polyaetiologic syndrome, which develops in people suffering from changes of the small intestine with the impairment of a process of absorption, that is, malabsorption syndrome and the following formation of immunoaller-gic reactions.

- The latter is revealed by the formation circulating immune complex with a deposition on reticulate fibers of dermal papillae Ig A and production of antireticular antibodies to a basal membrane and formation of subepidermal blisters.

Clinic.

- The disease may begin at any age but it is more common at the age of 30-40.
- It is characterized by the expressed seasonal occurrence (the improvement is in an autumn or winter).
- Usually the prodromal stage is gradual but in children it may be acute and is followed by subfibrilititis, malaise, dyspepsia and artralgia.

- In adults the disease may begin with subjective feelings (pricking, burning, itching) which may occur some hours or even days before the eruption appearance.
- A real polymorphism: simultaneous or appearance with some intervals of vesicles, papules, blisters on the limited erythematous area is charactestic for eruptions.

- The secondary morphological elements: crusts, erosions, excoriations appear after them.
- The eruptions are characterized by the grouped (the type of herpes) symmetrical location.
- General condition does not changed in spite of periodical rises of temperature.

- The favorite localization has not been observed but frequently eruptions are located on the skin of the trunk (lumbar – sacral region), buttocks, extensor surfaces of extremities. Pale-yellow blisters are typical for dermatosis at During of 1 cm in diameter.

- They may confluent into larger blisters with the following dryness and formation of crusts or quickly epithalizing erosions.
- The marked burning, itching and painfulness increasing by the evening follow eruptions.
- Mucous membranes are impaired considerably rarely than at vulgar pemphygus (10%), and are never the first symptoms of the disease.

- The patients have an increased sensitivity to halogens: iodine, bromide. So, in some doubtful cases **Jadasson test** is used (applications on the skin of 50 % ointment of iodine potassium).
- In the administration exacerbation of the process is observed. New, frequently blisters eruptions appear on the place of application in 24-48 hours.

Diagnostics.

- Diagnosis of herpertiformed During dermatosis is based on the presence of subepidermal blisters (revealed histological), characteristic clinic, eosinophilia in the blood and contents of the blisters, absence of symptoms of acantholysis (Nicklosky, Asbo-Hansen).
- By an immunofluorescent test in the area of the basal membrane Ig A depositions are revealed.

Treatment

- The treatment must be carried out on the background of whithoutgluten diet: to exclude wheat, rice, vats, rye, barley, millet and other cereals as well as products containing iodine (sea fish, sea cabbage, sausages and others), and also toothpaste, containing fluoride.

- Before the administration of treatment the correction of the alimentary tract is necessary on the base of duodenum function examination, the sanation of focal infection and especially the malignancy of the internal organs exclusion, because During dermatosis is a often a paraneoplastic marker.

- The main remedy for all age groups is administration of sulphones (diucyphon, dupson, avlosulphon and so on). Preparations are administered by cycles of 5-6 days
- But in case of a blisters variety of dermatosis sulphones may be ineffective. Glucocorticoids in dose of 20-40 mg of prednizolone during 2-3 weeks are administered additionally.

Congenital bullous epidermolysis

This disease is of genodermatoes origin, e.g. a congenital disease connected with the genes mutation aetiopathogesis should not factor is a genetically stipulated defect in the derma structure resulting in the increased activity of callagenaze, the elastic fibred are eliminated or absent.

- It is considered that the cause of the disease is the impairment of mucopolysaccharides exchange, their depolymerization, disturbance enzymes process and physicochemical balance. The impairment of immune homeostasis, immunosupression, in particular, had been established.

- Two main forms of the disease (they are about 20) are distinguished: simple and dystrophic.
- **Simple form** is a benign one. Most frequently it can be observed and had autosomnodominantly type of heredity. Boys are often ill.

- The clinical picture shows a very quick (some hours) development of blisters on the skin on the place of not large injuries, most often on the elbows knees, soles, legs, buttocks, hands, around the mouth in children of the breast age.

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- The blisters appear on the unchanged skin. They are superficial with transparent or yellowish (hemorrhagic) contents, 2-3 days later the blisters open and form erosion with a crust on it's surface.
- When the crust goes out the scars do not form, a slight pigmentation is possible.

- Nickolsky symptom is negative. The acantholytic cells are not revealed.
- Positive isomorphic reaction of Köbner is marked: on the place of friction new blisters appear.

- The mucous membranes are involved extremely rarely. General condition of the child does not usually change.
- By the period of a sex ripening the course of the disease considerably improves, many children have spontaneous recovery.

- **Dystrophic form**. Depending on the character of heredity hyperplasic and polydisplastic forms are distinguished.
- **Hyperplasic form** is inherited autosomnodo-minantly and is characterized by the spontaneous blisters appearance on the skin, rarely in the mucous membrane of the mouth at the age of 4-10 years.

- The scar atrophy is formed on the place of the blisters. The skin is dry, dystrophy of the nails, hyperkeratoses of the palms and soles, tooth anomalities are observed.

- **Polydisplastic form**. The heredity is autosomic-recessive. It is the severest form of the inherited bullous epidermolysis.
- Numerous large blisters with hemorrhagic contents (sometimes during the first days of life) appear spontaneously and are not connected with a mechanical impact.

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- Erosions appearing on their places are bleeding, painful; sometimes vegetations accompany them. Healing comes as scar atrophy.
- The blister may also appear on the mucous membrane of the mouth, esophagus, upper respiratory tract, conjunctiva and result in the stenosis of the larynx, esophagus, trachea, bronchi, keratitis with a proper several clinical picture.

- The skin is dry, atrophic, ancyloses, bone dystrophies up to the bony tissue of the hands and soles dissolvement mutilation – spontaneous amputation are observed.

- The patients usually die being children, but sometimes they live up to 25 years and die of general amyloidosis (deposition of amyloidal complex in the lungs, kidneys, liver and so on).

Treatment.

- There are no specific methods of treatment. General health remedies: ferrum preparations, calcium, j – globulin, blood transfusions, plasma transfusions, A.E. vitamins are used, at metabolic disturbances anabolic steroids (retabolil, nerabol), methyluracyl, pentoxile, levamisole and others are used.

- The most effective are combinations of diphenin, zink oxide and aevite which are used by courses during 1 month with intervals of 3-4 weeks.
- The positive effect, especially at a simple form is observed at administration of small doses of antimalaria drugs (chingamyn, chlorochin, delagil and so on).

- Large doses of corticosteroids in combination with antibiotics and antibacterial external remedies are used in lethal forms (severe polydysplastic manifestations).

- In case of local therapy the covering of the blister is pricked with a sterile needle, aniline colours are used, epithelizing and disinfecting external drugs (irucsol, vulnusan, curiasin and so on) are administered.