

Extramammary Paget's Disease

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What is Extramammary Paget's disease?

Paget's disease is a type of cancer that often affects the nipple. Extramammary Paget's disease (EMPD) refers to the same disease process. But, it occurs in other areas of the body that are **not** the breast. These areas include the vulva, the skin around the anus, the armpits, groin, ear canal, and eyelids. The areas where this disease occurs have a lot of apocrine glands. Apocrine glands secrete things like sweat.

Extramammary Paget's Disease makes up 6.5% of all cases of Paget's disease. Extramammary Paget's disease can occur as a **primary process**. This means that the cancer starts in the affected area. It can also occur as a **secondary process**. This means that the cancer started somewhere else in your body and spread to the skin. This occurs in 35-40% of all cases.

How do doctors know I have Extramammary Paget's Disease?

If your doctor suspects you have this disease, they will perform a biopsy. This means that they will take a piece of the skin tissue. Another doctor will then look at this tissue under a microscope to confirm the diagnosis.

Your doctor may also use special stains to help confirm the disease. These are called immunostains. Immunostains stain certain proteins. All cells have proteins on their surface. Some cells have different proteins than other cells. The special stains help doctors figure out the types of cells they see under the microscope. This can be helpful to confirm the diagnosis of Extramammary Paget's Disease. It can also help them tell if this is a primary or a secondary process.

What does Paget's disease look like under the microscope?

Paget's disease and extramammary Paget's disease have cancer cells in the lower portions of the epidermis. This can make the basal cell layer look "crushed". The cancer cells often have pale cytoplasm. They also have pleomorphic nuclei. This means that the nucleus of the cancer cells vary in size and shape.

As the cancer progresses, tumor cells may be seen moving up toward the upper layers of the epidermis. But, most of the cancer cells will still be just above the basal layer. Cancer cells in Extramammary Paget's Disease have a lot of mucin.

Paget's disease can look like other cancers that affect the epidermis. These include:

- Bowen's disease. This is a type of squamous cell carcinoma.
- Melanoma.

All these diseases may have cells moving up toward the top of the epidermis. This is known as "pagetoid spread". For this reason, immunostains are needed to confirm the disease.

What are the immunohistochemical staining patterns of Extramammary Paget's disease?

Extramammary Paget's Disease will stain positive for CK7, CEA, and PAS.

Doctors can rule out melanoma as the cancer cells will be negative for melanocyte markers, like S100. Melanocytes are cells where melanomas arise from.

Doctors can rule out squamous cell carcinoma as the cancer cells will be negative for keratin and positive for CEA.

What is the outcome of Extramammary Paget's Disease?

The outcome of this disease is fairly good. But, higher rates of death occur in Extramammary Paget's Disease that is a secondary process.

It is fairly common for this cancer to recur at the affected area after treatment. But, it is rare for the cancer to spread to distant organs if it is a primary process. It can spread to the lymph nodes in some cases.

Factors that affect outcomes include:

• Nodules in the affected area of the skin • High levels of CEA	• If the cells are found deep in the tissue under the microscope • If the cells spread to the lymph nodes
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