In 1874 Sir James Paget first described the disease of the nipple that 15 years later became known as mammary Paget's disease. This was followed by reports of extramammary Paget's disease (EMPD) in areas such as the scrotum, penis, and vulva.

EMPD is an unusual intraepithelial adenocarcinoma of unknown etiology that occurs primarily in apocrine gland-bearing areas of the body. It is not analogous to mammary Paget's disease, which is an adenocarcinoma that arises in large lactiferous ducts (usually the nipple) and extends into the epidermis. The most common site for EMPD is the vulva, followed by the anogenital area in men. It typically occurs in white men and women between the ages of 50 and 65 and is nearly 5 times more prevalent in women.

EMPD usually begins insidiously with complaints of pruritus and a burning sensation. Clinically, the appearance of the lesion ranges from scaling and flaking to oozing and maceration; it may even evolve to a crusted plaque and tumor formation. Delay in diagnosis is common, as many cases are erroneously treated initially as fungal infections, candidiasis, psoriasis, or eczematous eruptions. The extent of the lesion closely correlates with the duration of symptoms preceding surgery. Therefore, any rash in the anogenital area that is not responsive in 6 to 8 weeks to topical therapy should be biopsied to exclude EMPD.

The diagnosis is confirmed by a histologic examination that reveals Paget's cells in the epidermis. EMPD may become invasive and metastasize via the lymphatic system, and is associated with an underlying malignancy in 14% to 20% of patients (the bladder and rectum are the most common sites) or an adnexal adenocarcinoma in 4% to 7% of patients. Therefore, a systemic evaluation is required.

Surgical excision is considered the standard treatment. Invasive tumors may require extensive surgery with lymph node dissections.

For in situ lesions the prognosis is typically good; however, multiple excisions may be needed, because local recurrences are frequent. With invasion, despite the extent, the metastatic rate is high. Patients with associated carcinomas of contiguous visceral organs, either genitourinary or gastrointestinal, have a poor prognosis. Therefore, long-term follow-up is necessary.

**References**


The complete differential diagnosis and the pathologic discussion are available at [www.JHASIM.com](http://www.JHASIM.com).