How do I diagnose *Pagetoid Bowen Disease*

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**Differential diagnoses:** Paget Disease, Malignant Melanoma in situ

**Side findings:** subepithelial lymphoplasmacytic infiltrate, elastotic degeneration.

**Abstract**

Bowen disease is a variant of squamous carcinoma in situ, which sometimes presents with a pagetoid growth pattern with cytologically atypical clear cells. In that case, the differential diagnosis includes Paget disease and melanoma in situ, as well as less common entities. Here we describe a case where histological differentiation is difficult and immunohistochemical studies are necessary to make a diagnosis. Here we also highlight the heterogeneity of CK7 expression and the apparent specific expression of p63 in Pagetoid Bowen Disease.

**Virtual Slides:** [www.diagnosticpathology.eu/vs/2015_1_88/](http://www.diagnosticpathology.eu/vs/2015_1_88/)

**Anamnesis / History**

The patient was an 88 year-old male, who had undergone excision of a skin tumor of the left malar region. He also had a history of adenocarcinoma of the colon.

**Gross - microscopic findings**

We received a 26x17x7mm skin ellipse. There was a 18x10mm white, irregular and ulcerated plaque at the epidermal surface. The closest margin was 3mm from the lesion.

**Microscopy**

Microscopy revealed cytologic atypia of all levels of the epidermis, with an intraepithelial proliferation of neoplastic cells with atypical nuclei (with prominent nucleoli) and pale vacuolated cytoplasm. These cells had a pagetoid distribution, in not well demarcated nests or in single cells. Although the neoplastic cells extended to the
basement membrane, they did not flatten the basal layer. Extension to the follicular epithelium at the level of the sebaceous duct was also identified. There was focal hyperkeratosis and parakeratosis of the surface epithelium, and also areas of ulceration. We also observed subepithelial lymphoplasmacytic infiltrate and elastotic degeneration of the superficial dermis.

**Expression of markers**
CK7(pos), p63(pos), CEA(neg), Melan A(neg).

**Discussion**
A pagetoid neoplastic infiltration of the epidermis encompasses a differential diagnosis that includes primarily Paget Disease, Melanoma in situ and Bowen disease. Histological differentiation of these entities can in some cases be difficult and use of immunohistochemical markers maybe necessary to provide a definitive diagnosis. Melan A is a marker for melanoma, which this lesion did not express, excluding this diagnosis. Usually, CK7 and CEA are thought to differentiate Paget Disease from Pagetoid Bowen Disease, since they are expressed in Paget Disease and almost invariably negative in Bowen Disease. However, there have been reported cases of Pagetoid Bowen Disease expressing CK7 and CEA. Another proposed marker for this differentiation is p63, which is expressed by the stratified epithelia and as so is thought to be specific for Bowen Disease. In this case, the lesion did not express CEA but it did express CK7 and p63, the latter being the definitive diagnostic marker.

**Hallmarks of Diagnosis**
Pagetoid distribution pattern of neoplastic cells, expression of p63.

**Images** (for full size images see supplements)
Keyword - Diagnosis: Pagetoid Bowen Disease
Keyword - differential diagnosis: Paget Disease
Keyword - side findings: elastotic degeneration
Keyword - organ: Skin
Keyword - methods: p63

Online References (PubMed)