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## Grzybowski generalized eruptive keratoacanthoma

Grzybowski syndrome, or generalized eruptive keratoacanthoma, refers to a very rare disease in which hundreds of keratoacanthoma-like papules appear. The cause is unknown. It should be distinguished from an inherited disorder, Fergusson Smith familial keratoacanthomas, in which multiple keratoacanthomas appear in children and young adults.

### Clinical features of Grzybowski syndrome

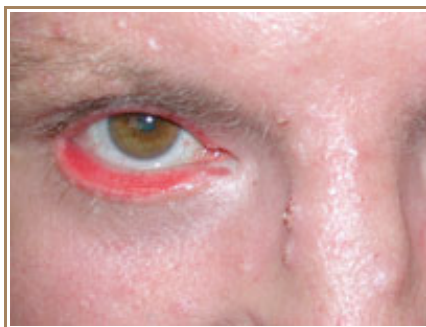
Only about 30 cases of Grzybowski's syndrome have been written up in the medical journals worldwide. The syndrome affects middle-aged adult males and females equally. It starts abruptly and extends over some months. Untreated, it is reported to persist indefinitely. The main features are:

- Severe itch
- Numerous firm bumps on all affected areas (face, scalp, trunk and limbs)
- White bumps in the mouth (lips, tongue, inside the cheeks)
- Larger horny nodules that persist for a few weeks, then disappear by themselves
- Koebnerisation – the nodules may appear at sites of skin injury
- Mask-like facial appearance, due to thickened face skin
- Ectropion (drooping lower eyelids) and retraction of eyelid skin
- Hoarseness due to nodules on the larynx (voice box)

#### Grzybowski syndrome



Oral and perioral papules



Eyelid retraction



Diffuse eruption on chest



Eruptive keratoacanthomas



Eruptive keratoacanthomas



Diffuse rash

## What is a keratoacanthoma?

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The papules and nodules seen in Grzybowski syndrome resemble a common skin lesion, [keratoacanthoma](#). However, keratoacanthoma is derived from hair follicle cells and does not normally affect mucosal surfaces such as the mouth as is the case in Grzybowski syndrome. Keratoacanthoma may favour sun exposed areas, but can affect covered areas as well. Like keratoacanthoma, individual Grzybowski lesions erupt, persist for a few weeks and then clear.

Witten and Zak described a similar condition in which there were giant keratoacanthomas.

Luckily, despite the resemblance of the eruptive keratoacanthomas to the skin cancer, [squamous cell carcinoma](#), they behave in a benign fashion in Grzybowski syndrome. The lesions do not spread to the lymph nodes or affect internal organs so general health is unaffected. However, the severe itching and abnormal appearance can lead to severe depression.

## Investigations

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Most individuals with Grzybowski syndrome are otherwise in good health, and blood test results are entirely normal.

Skin biopsy of Grzybowski lesions shows similar changes to keratoacanthoma.

- Keratin-filled core
- Thickened squamous layer of the skin
- Atypical skin cells (keratinocytes)
- Proliferation of keratinocytes extending into the dermis (lower layer of skin)
- Mild inflammation

Because in some ways these lesions look and behave like [viral warts](#), several investigators have looked for evidence for human papillomavirus within the lesions. The results have been inconclusive, but in most cases no evidence of viral infection has been found.

## Treatment

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Unfortunately, treatment fails for many patients with Grzybowski syndrome. Only one case has been reported to clear up spontaneously. In other cases, improvement or complete resolution has occurred with one of the following medications:

- [Acitretin](#)
- [Isotretinoin](#)
- Fluorouracil
- [Methotrexate](#)
- [Cyclophosphamide](#)

Symptoms can be eased with [emollients](#) and anti-itch medications.

### Related information

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#### On DermNet NZ:

- [Keratoacanthoma](#)
- [Skin lesions](#)

#### Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.  
If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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