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Paraneoplastic Pemphigus

By Sally Robertson, BSc

Pemphigus diseases are a rare group of autoimmune conditions characterized by the formation of blisters that affect the skin and / or mucous membranes. Paraneoplastic pemphigus (PNP) is a rare form of pemphigus that is generally associated with lymphoproliferative disorders such as non-Hodgkin's lymphoma, Castleman disease, and chronic lymphocytic leukemia. More rarely, the condition is associated with non-lymphoid cancerous neoplasms. PNP is the most rare and most serious form of pemphigus, especially due to the underlying malignancy in affected individuals.

Antigens from the tumor stimulate immune responses that lead to the formation of blisters on the mucous membranes and other epithelial membranes. The condition often affects the respiratory tract and gastrointestinal tract, with painful erosions usually forming on the lips and in the esophagus and mouth. Skin eruptions may also occur and eventually form blisters.

Who is Affected?

PNP can affect people of any race or gender equally. The condition can develop in children but the average age of disease onset is 60 years.

Symptoms

- Oral lesions: The oral erosions that develop are usually severe and result in crusting on the vermillion of the lips.
- Skin lesions: The skin lesions are highly variable and may be confused with other conditions that involve the formation of blisters such as pemphigus vulgaris and erythema multiforme. Examples of the types of lesions that affect the skin include papules, bullae, scaly plaques, erosions, and erythroderma.
- Respiratory lesions: Unlike with other forms of pemphigus, PNP affects the lungs. It leads to constrictive bronchiolitis and generally causes irreversible damage to the lungs that significantly increases the risk of mortality.
- Other lesions: Other surfaces that may be affected by lesions include the genitalia and the inside of the nose.

Diagnosis of PNP

Since PNP is an IgG-mediated disease, a blood test is usually performed during diagnosis to check for the presence of IgG antibodies. Two types of skin biopsy are also performed. A blister is removed and sent for histological analysis and a biopsy of the skin within 2cm of the blister (peri-lesional skin) is taken and sent for direct immunofluorescence testing (DIF). In the case of a positive diagnosis, histology shows acantholytic cells inside blisters and dead keratinocytes. The DIF reveals IgG antibodies that have targeted proteins that form the desmosome complex, a structure required for cell-to-cell adhesion.

Management of PNP

The underlying malignancy needs to be identified and treated for PNP to resolve. Pulmonary disease is treated with potent immunosuppressants such as azothioprine or cyclosporin. In some cases, plasmapheresis may also be performed. Other treatment approaches are generally supportive such as dressing the skin, preventing secondary infections, and administering pain relief.

Prognosis

This multiorgan syndrome is generally associated with poor patient outcome and high mortality, with around 80% of patients dying as a result of PNP or associated malignancy. The average survival time is less than one year and the main causes of mortality are development of the malignancy, respiratory failure, and sepsis.

Reviewed by Susha Cheriyedath, MSc

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Further Reading

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