



Case Report

Fine-needle aspiration cytology – A novel diagnostic technique in pemphigus

Beena Narayanan¹, O. T. Dhanya², N. Asokan³

¹Department of Dermatology, Government Medical College, Kottayam, ²Department of Dermatology, Primary Health Center, Wayanad, ³Department of Dermatology and Venereology, Government Medical College, Thrissur, Kerala, India.

***Corresponding author:**

Beena Narayanan,
Professor and Former Head,
Department of Dermatology,
Government Medical College,
Kottayam, Kerala, India.

drbeenanarayanan@yahoo.co.in

Received : 04 December 2020

Accepted : 14 December 2020

Published : 06 April 2021

DOI:

10.25259/JSSTD_59_2020

Quick Response Code:



ABSTRACT

The term pemphigus is collectively used to describe a group of immunobullous disorders characterized by intraepidermal cleft and circulating antibodies against intercellular adhesion molecules. Pemphigus vegetans was first described as a variant of pemphigus vulgaris by Neumann in 1876. We report a case of 36-year-old female who presented with swelling of scalp, which was diagnosed as pemphigus by fine-needle aspiration cytology.

Keywords: Pemphigus vegetans, Diagnosis, Fine-needle aspiration cytology

INTRODUCTION

Pemphigus is a group of autoimmune blistering disorders with intraepidermal cleft caused by immunoglobulins directed against keratinocyte cell surface antigens.^[1] Pemphigus vegetans (PVe) is a rare variant of pemphigus vulgaris (PVu) which commonly presents with vegetating lesions on the flexural regions, face, and scalp and mucosal erosions. The disorder affects chiefly middle-aged adults.^[2-5] We report a case of PVe presenting with a firm swelling on the scalp which was diagnosed as pemphigus by fine-needle aspiration cytology (FNAC).

CASE REPORT

A 36-year-old lady presented with pustular lesions on the left big toe, axillae, groins, and face of 1 month duration, and an asymptomatic smooth swelling on the scalp, which was gradually increasing in size. Physical examination revealed well-defined erythematous plaques of size 5 × 4 cm, with pustules, erosions, slough, and crusting below the left eye and the left big toe [Figure 1a] and vegetating plaques in the axillae [Figure 1b] and groins. There was a well-defined, firm, non-tender swelling on the vertex of the scalp of size 16 × 10 cm without any hair loss. Oral mucosa showed multiple erosions. The patient was investigated with a differential diagnosis of pyoderma gangrenosum and PVe. Pathergy test was negative. Tzanck smear from the pustules revealed numerous acantholytic cells and eosinophils. Histopathology showed hyperkeratosis, acanthosis, suprabasal cleft with acantholytic cells, and dermal inflammatory infiltrate composed of eosinophils, neutrophils, and lymphocytes [Figure 2].

Direct immunofluorescence (DIF) of perilesional skin showed intercellular staining (ICS) of epidermis with IgG and C3 [Figure 3]. Indirect immunofluorescence (IIF) showed ICS with

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IgG in 1:10 titer. Although we could confirm the diagnosis of PVe (Hallopeau type), the cause of the scalp swelling was obscure. We, therefore, proceeded with ultrasonography of the scalp swelling and FNAC from it. To our surprise, FNAC demonstrated plenty of acantholytic cells and eosinophils

[Figure 4]. This confirmed that the scalp swelling was a component of PVe. The patient was treated with parenteral dexamethasone (initiated at 4 mg twice a day); swelling of the scalp flattened [Figure 5] and subsided in a week, leaving no residual changes. Other lesions also resolved in a month's time.



Figure 1: (a) Erythematous plaques with pustules, erosions, and slough on the left big toe of a patient with pemphigus vegetans. (b) Vegetative plaques in the axilla.

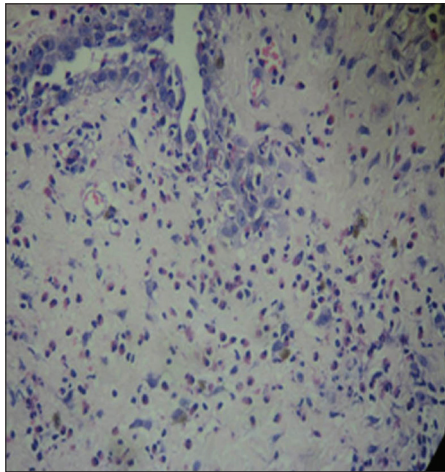


Figure 2: Histopathology showing suprabasal cleft with acantholytic cells and eosinophils. Dermis showing eosinophils and neutrophils (H and E, x400).

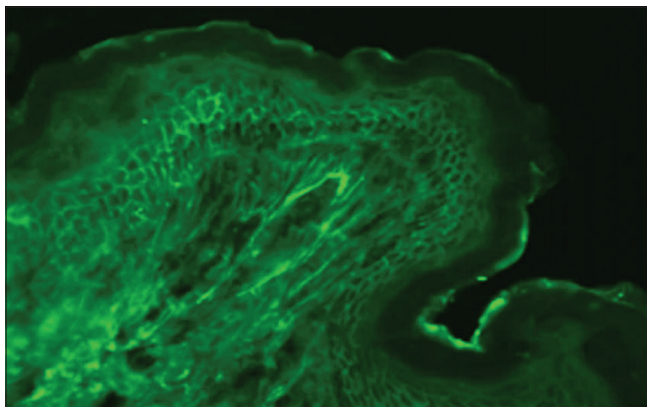


Figure 3: Direct immunofluorescence on perilesional skin of a patient with pemphigus vegetans showing intercellular staining with IgG and C3.

DISCUSSION

PVe constitutes about 1%–2% of all cases of pemphigus. The principal autoantigen in Pve is desmoglein 3. Other antigens identified are desmoglein 1 and desmocollins.^[5] Historically, PVe has been divided into two subtypes – Neumann and Hallopeau.^[6,7] In Neumann type, the lesions begin as vesicles and bullae that evolve into vegetating masses which bleed

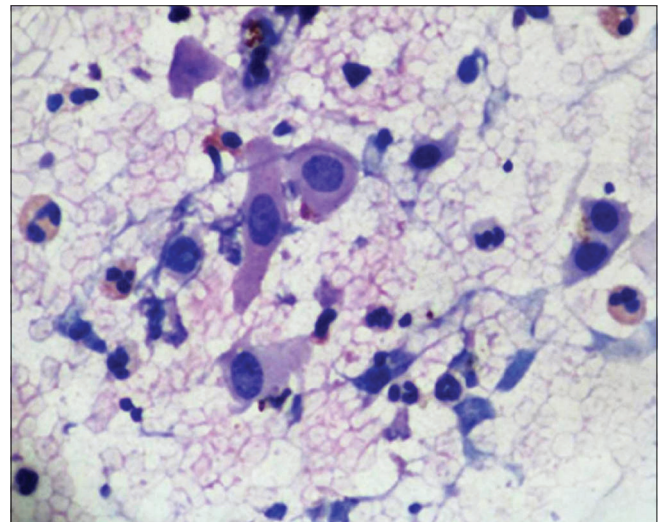


Figure 4: Fine-needle aspiration cytology from a swelling on the scalp of a patient with pemphigus vegetans showing acantholytic cells (Papanicolaou stain, x400).



Figure 5: Swelling of the scalp showing flattening after administration of systemic corticosteroids.

easily on trauma.^[6] The disease is similar to PVu in relapses, remissions, response to therapy, and prognosis.^[6] Hallopeau type is relatively benign. Pustules are the primary lesions which gradually progress to vegetating plaques. Spontaneous healing and remissions may occur.^[6,8] Oral lesions are present in over 90% of the patients.^[6] Scalp lesion is common in PVu but not a classic feature of PVe.^[9]

Histopathology shows hyperkeratosis, papillomatosis, acanthosis with plenty of eosinophils, and acantholytic cells in the suprabasal clefts and a heavy infiltrate of eosinophils, lymphocytes, and neutrophils in the dermis.^[10] DIF of perilesional skin demonstrates epidermal intercellular IgG, sometimes with C3.^[2,4,5,6,11] IIF demonstrates circulating intercellular antibodies in many patients.^[11,12] FNAC is proved useful in diagnosing various benign and malignant skin conditions, but to the best of our knowledge, is not yet utilized in diagnosing pemphigus.^[13]

CONCLUSION

We, suggest that FNAC can be added to the diagnostic armamentarium in pemphigus, as a simple, fast, and reliable technique.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

Dr. Beena Narayanan and Dr. N Asokan are on the editorial board of the Journal.

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How to cite this article: Narayanan B, Dhanya OT, Asokan N. Fine-needle aspiration cytology – A novel diagnostic technique in pemphigus. *J Skin Sex Transm Dis* 2021;3(1):77-9.