

Clinicopathological and Demographic Characteristics of Paget's Disease: a 4-year Study Showing a Male Predominance in Extra-mammary Paget

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Introduction

Paget disease is a rare skin neoplasm and is categorized into mammary (MPD) and extra-mammary (EMPD) [1,2]. To date, its detailed histopathological features along with patients demographic characteristics have not been investigated in Iran. In this study, we performed a clinic-pathological analysis of Paget cases in a single tertiary center between 2016 and 2020.

Cases Presentation

A total of 32 cases including 26 (81.25%) females and 6 (18.75%) males with the mean age of 53 ± 9.96 years

(range 34-78) were included in our study. All patients gave informed consent for participating in our study. The majority of our patients 26/32 (81.25%) had MPD, and 6/32 (8.75%) had EMPD. All cases had a history of scaly and erythematous prolonged lesions. Out of the 26 MPD cases, 25/26 (96.1%) were female and 1/26 (3.9%) were male. The mean age of the MPD cases was 51.03 ± 8.80 (range 34-78). Out of the 6 EMPD cases, 5/6 (83.3%) were male and 1/6 (16.7%) were female. The mean age of the EMPD cases was 61.5 ± 10.98 (range 42-74). Of note, all EMPD patients had ano-genital involvement. 2/6 (33.33%) cases had scrotum EMPD, 1/6 (16.66%) patient had penis and scrotum EMPD, 2/6 (33.3%) had peri-anal involvements, and

Table 1. The results of immunohistochemistry staining.

	Number of patients that were assessed for the presence of this antigen	Number of patients that were positive N (%)
CK7	6	6 (100%)
CK AE1/AE3	6	6 (100%)
S100	3	2 (66.66%)
CEA	6	5 (83.4%)
Melan A	4	3 (75%)
Ki-67	2	2 (100%)
HMB-45	2	1 (50%)

1/6 (16.66%) patient had labia major EMPD. Histopathological examination revealed the presence of cells with pale cytoplasm in addition to prominent nuclei along with a high nuclei cytoplasm ratio (N/C) in all 32 cases. Moreover, 10/32 (31.25%) had lymphocyte infiltration. IHC was performed for 6 EMPD patients and all of them were positive for CK7 and (CK AE3/AE1) (Table 1).

Conclusions

It is believed that Paget disease is more prevalent among older female patients [1-5]. The peak incidence of MPD is reported after the age of 60, while the mean age of the MPD patients in our study was 51 years [1].

Asian studies have shown a significant EMPD predominance in males and likewise, in our study, we found that the majority of EMPD patients were men, although, other studies have reported a higher prevalence of EMPD in female patients [1]. The most common location for EMPD is the vulva in 65% of cases, perianal area, scrotum, penis, and axilla [1]. In this regard, the only female EMPD patient in our study had labia major involvement. Based on the Ghazawi et al study, the most common sites for EMPD in males are the scrotum and penis, which is consistent with the findings of our study [4].

The histologic hallmark of Paget disease is the presence of Paget's cells [1]. In our study, the pathologic examination revealed the presence of Paget cells in all cases along with lymphocytic infiltrations in one-third of patients. In a study done by Elbendary et al the histopathologic evaluation showed no significant differences between MPD and EMPD which is consistent with our study [5].

Our study confirms that MPD is more prevalent than EMPD. Based on our study, there is a female predominance in MPD cases at younger ages in comparison to other reports, and penis and scrotal involvements of EMPD are highly probable locations.

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