

The Epidemiology and Clinical Characteristics of Extramammary Paget Disease Patients in Canada and Assessing the Risk of Second Malignancies



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Abstract

Extramammary Paget Disease (EMPD) is a rare intraepithelial adenocarcinoma that arises in apocrine gland-bearing skin and presents as a chronic rash that resembles several disorders such as eczema, cutaneous T cell lymphoma, Langerhans cells histiocytosis, intraepithelial neoplasia, fungal infections and psoriasis. EMPD is often associated with underlying or distant malignancies. The epidemiology of EMPD in Canada is incompletely understood. Further, the risk of developing second malignancies in Canadian EMPD patients was not evaluated prior to the presented study. In this study, we thoroughly analyzed clinical characteristic, incidence and geographical distribution of EMPD patients in Canada from 1992 to 2015 using the Canadian Cancer Registry (CCR). In summary, in Canada, 845 patients were diagnosed with EMPD between 1992 and 2015. The mean incidence rate of EMPD in Canada was 1.1 cases per 1 million individuals per year with noteworthy variations in the incidence across the country. The male:female incidence ratio of EMPD was 1.00:1.87. The incidence of EMPD in Canada increased steadily within the aforementioned period. Analysis of the occurrence of second malignancies in EMPD patients revealed that 29.6% of EMPD patient developed at least one additional malignancy. These malignancies were predominantly adenocarcinoma at gastrointestinal sites. In conclusion, this study describes the epidemiology and patient distribution of EMPD patients in Canada. Further, the study also confirms increased risks of second malignancies in EMPD patients. Epidemiological studies, such as the presented one here, shed light on the risk factors associated with the development of EMPD and encourage regular screening of EMPD patients for second malignancies.

Background

- EMPD is a rare intraepithelial adenocarcinoma that arises in apocrine gland-bearing skin.
- It presents as a chronic rash that resembles several conditions including eczema, Bowen's disease, mycosis fungoides, Langerhans cells histiocytosis, intraepithelial neoplasia, fungal infections, psoriasis, etc.
- Affects men and women and originates in the perianal and genital regions, as well as other skin sites.
- Pruritus and pain associated with an eczematous, erythematous, or ulcerated rash in individuals aged 50–80 years is the typical presentation.



- The exact pathogenesis of the disease is unclear; however, current evidence suggests that malignant transformation of intraepithelial pluripotent cells gives rise to adenocarcinoma.
- Due to the scarcity of EMPD cases, only few case-series and few population-based studies examined the epidemiology of this disease globally.
- Reported incidences of this disease are 0.6 cases per million person years in Europe and in the United States it ranges from 0.3 in Blacks, to 0.9 in Whites to 3.7 in Asians.
- Little is known on epidemiology of EMPD patients in Canada.
- EMPD has been shown in multiple studies to be associated with increased risk of a secondary cancer, predominantly adenocarcinoma. This was never investigated in Canada using a population-based database.

Objectives

- To investigate the epidemiology of EMPD patients in Canada.**
- To investigate the demographic characteristics of Canadian and EMPD patients.**
- To investigate the risk of secondary malignancies in EMPD patients this patient population.**

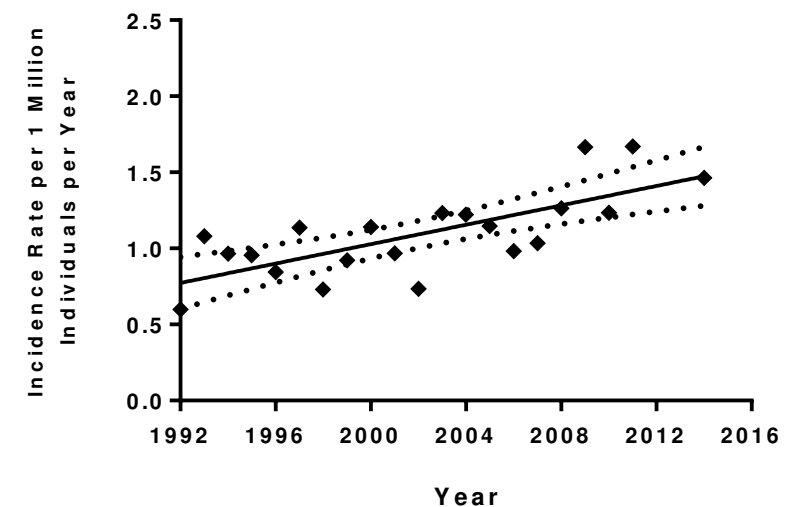
Methods

- Two Canadian population-based Cancer registries were examined: Canadian Cancer Registry & Régie de l'assurance maladie du Québec.
- Clinical characteristics (age, sex, anatomical location) of EMPD patients were analyzed.
- Additional data from EMPD patients were extracted to determine the occurrence of secondary malignancies.

Results

- 845 Canadian patients were diagnosed with EMPD.
- The mean age of diagnosis = 71.13 years.
- The mean incidence rate of EMPD in Canada was 1.1 cases per 1 million individuals per year.
- Overall incidence of EMPD was significantly higher in females. The male:female incidence rate ratio of EMPD in Canada is 1.0:1.87

Incidence trends of EMPD in Canada



Incidence trends of EMPD in Canada. The mean incidence rate of EMPD in Canada was 1.1 cases per 1 million individuals per year.

Secondary malignancies in Canadian EMPD patients

- Out of 845 Canadian patients who were diagnosed with EMPD, 250 patients developed additional malignancies (before or after EMPD diagnosis) = **29.5%**
- These malignancies were predominantly adenocarcinoma at gastrointestinal sites, prostate, breast and lungs.
- The mean time between EMPD and the cancer before and after EMPD was 6.41 and 6.58 years, respectively.

Conclusion

- The study confirms increased risks of second malignancies in EMPD patients.
- Epidemiological studies, such as the presented one here, encourage regular/interval screening of EMPD patients for additional malignancies (colonoscopy, FOBT, mammograms, chest X ray, etc).

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