Prevalence of Extramammary Paget's Disease in Urban China: A Population-based Study

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Abstract

Background

Extramammary Paget's disease (EMPD) is an intraepithelial adenocarcinoma. The chronic relapsing clinical course and unbearable clinical symptoms of EMPD usually result in a markedly diminished quality of life. No national data are available on descriptive epidemiology of EMPD in China, the most populous country over the world. This population-based study aimed to estimate the prevalence and associated sex and age patterns of EMPD in China.

Methods

This study was conducted using data from China's Urban Employee Basic Medical Insurance and Urban Resident Basic Medical Insurance, covering approximately 0.43 billion Chinese urban residents in 2016. Patients with EMPD were identified based on the diagnostic names and codes in claim data.

Results

A total of 53 males and 31 females with EMPD were found. The crude prevalence in 2016 was 0.04 per 100 000 population [95% confidence interval (CI): 0.02 to 0.06], ranging from 0.01 (95% CI: 0.00 to 0.02) in North or Northeast China to 0.08 (95% CI: 0.03 to 0.16) in Southwest China. The rates were higher in males (0.05, 95% CI: 0.03 to 0.08) compared with females (0.03, 95% CI: 0.02 to 0.05). The mean age of patients was 65.87 (standard deviation: 14.21) years, with the peak prevalence appeared in patients aged 70–79 (0.28, 95% CI: 0.16 to 0.42).

Conclusions

The prevalence of EMPD was markedly lower than those in the United States and Europe, and varied across regions in China. Chinese patients were much younger, with significant male predominance. Further study is warranted to examine potential pathophysiologic mechanism.

Background

Extramammary Paget's disease (EMPD) is an intraepithelial adenocarcinoma which predominantly affects areas with a high apocrine gland concentration, including the vulva, perineal, perianal, scrotal, penile and axillae skin.[1–3] EMPD lesions are notorious for its chronic relapsing clinical course and unbearable clinical symptoms such as itching, skin ulcer and bleeding.[2, 4] The standard treatment for EMPD is often extensive surgical excision followed by plastic reconstruction, which frequently leaves anatomical and functional impairments, and results in a significantly diminished quality of life.[4, 5] Understanding the epidemiological characteristics of EMPD is critical to inform policy making about

EMPD management. However, epidemiological information of EMPD was limited, especially in the developing countries including China.

EMPD was reported to be most common in individuals between 60 and 80 years of age, with a peak incidence at 65 years.[2, 6–8] A female predominance was reported in aged Caucasians, with a male to female ratio between 1:1.6 and 1:3.6 in western literatures.[9–13] By contrast, a male predominance was found by both single- and multicenter studies in China, Japan, and Korea, with a male to female ratio between 2:1 and 5:1.[14–17] The mean incidence rate of EMPD ranged from 0.054 patients per 100 000 person-years to 0.11 patients per 100 000 person-years in Europe.[12, 13, 18] In multiracial country like the United States, the incidence of EMPD showed ethnic disparity, the incidence in black males was nearly four times lower and in Asians/Pacific islander males four times higher than that in Caucasian males between 1973 and 2009.[1] No national epidemiological studies are available to estimate the prevalence and incidence of EMPD in Asian countries, including China.

This study was conducted to provide recent estimates of the prevalence of EMPD in mainland China and to investigate its patterns across sexes, age groups and geographical regions, using a nationally representative data in 2016.

Materials And Methods

Data sources

The anonymous claim data we used were from Urban Employee Basic Medical Insurance (UEBMI) and Urban Residence Basic Medical Insurance (URBMI). As two main health insurance schemes in urban China, UEBMI is oriented to working or retired employees and URBMI is for urban residents unemployed. By 2016, UEBMI and URBMI have covered approximately 95% of the urban population in China. Every city updated the data in both databases on a monthly basis. The reimbursement records of the insured population will be recorded in the database, no matter the proportion they paid for the medical service. Alongside the fees and procedures claimed, diagnostic information (i.e., disease names and disease codes) and sociodemographic characteristics (i.e., ethnicity, sex, birth date, place of residence, etc.) are recorded as well.

Study population

This national population-based study used the UEBMI and URBMI data of 23 provinces between January 1st, 2016 and December 31st, 2016. Eight provinces including Beijing, Shanghai, Sichuan, Ningxia, Hebei, Tianjin, Fujian and Tibet were excluded due to absence or abnormality of key information such as diagnostic information, only containing a single insurance type or reporting policy exemptions. The study protocol was approved by the ethical review committee of the Peking University Health Science Center (IRB. No.: IRB00001052-18012), and the informed consent requirement was waived.

Case identification

The identification of patients with EMPD was based on the diagnostic information in the database, such as diagnostic text and International Classification of Diseases (ICD) code. Natural language processing was utilized to normalize the diagnostic information with a dictionary of potential EMPD defined by prestigious clinicians. Potential patients with EMPD were selected by ICD-10 (C51.902, C50.903, M85400/3, C63.252, M85420/3, M85420/6) and Chinese medical terms of diseases including EMPD, Paget's disease of perineum, Paget's disease of pubic caruncle, Paget's disease of scrotum, Paget's disease of penis, extramammary eczematoid carcinoma, extramammary eczematoid carcinoma of perineum, extramammary eczematoid carcinoma of pubic caruncle, extramammary eczematoid carcinoma of scrotum and extramammary eczematoid carcinoma of penis. Two prestigious clinicians read the diagnostic information of each potential patient with EMPD independently to identify actual target patients. Any disagreements between them would be judged by another senior clinician.

Statistical analysis

Through a two-stage approach, the national prevalence of EMPD in 2016 was calculated. In the first stage, the prevalence in each province was estimated. In the second stage, we pooled the prevalence of each province using a random effects meta-analysis to calculate the national or regional prevalence. The details can be found in our previous research.[19]

The prevalence of EMPD was estimated by sex, age, and geographical region (East, North, Northeast, Northwest, Southcentral, and Southwest) as well. To test the robustness of the main results, sensitivity analyses were conducted by setting a stricter algorithm (not considering the patients with diagnostic information containing eczematoid carcinoma) to identify actual patients with EMPD, by only considering the observed patients with EMPD to estimate the lower bound of the prevalence, and by excluding the top 10% of provinces with missing diagnostic information. Based on the 2010 Chinese national census data, the Revised European Standard Population (RESP) 2013, the 2010 US population and the 2011 Australian population, age-adjusted rates of EMPD prevalence were provided. Prevalence with 95% confidence interval (CI) was estimated by Poisson distribution. Student's t-test was used for continuous variables and the chi-squared test was used for categorical variables. All statistical analyses were done by Stata version 15.0, and a 2-sided $P < 0.05$ was considered as statistically significant.

Results

Approximately 0.43 billion subjects were included in this study (Table 1). There were 84 patients with EMPD in the database in total (Table 2). The mean age of the observed patients with EMPD was 65.87 [standard deviation (SD):14.21] years. There were no statistically significant differences in mean age and area distribution between male and female patients with EMPD.

Prevalence

The 2016 prevalence of EMPD in urban China in 2016 was 0.04 per 100 000 population (95% CI: 0.02 to 0.06). Based on the Chinese census data in 2016, there were 0.79 billion urban population, therefore in

2016, approximately 316 patients with EMPD in China. Male prevalence (0.05, 95% CI: 0.03 to 0.08) was higher than the female prevalence (0.03, 95% CI: 0.02 to 0.05) (Fig. 1.). In terms of the age trend of EMPD prevalence, the prevalence reached a peak at 70-79 years old (0.28, 95% CI: 0.16 to 0.42) (Fig. 1.). The EMPD prevalence in different regions varied, with the rates in Northwest China (0.07, 95% CI: 0.02 to 0.16), Southcentral China (0.06, 95% CI: 0.02 to 0.11) and Southwest China (0.08, 95% CI: 0.03 to 0.16) were higher than other regions (Table 3 & Table 4).

Adjusted prevalence

The age-adjusted national prevalence based on 2010 Chinese census data was 0.04 per 100 000 population (95% CI: 0.04 to 0.05). The standardized prevalence rates by the US, European and Australian populations were 0.05 (95% CI: 0.05 to 0.06), 0.07 (95% CI: 0.06 to 0.08) and 0.06 (95% CI: 0.05 to 0.06), respectively.

Sensitivity analysis

The lower bound of the national prevalence was calculated as 0.03 per 100 000 population (95% CI: 0.02 to 0.04) by only considering the observed patients. The prevalence rates calculated by setting a stricter algorithm to identify patients with EMPD or excluding the top 10% of provinces with missing diagnostic information (i.e., Shandong and Xinjiang) were 0.04 (95% CI: 0.02 to 0.06) and 0.05 (95% CI: 0.03 to 0.07), respectively.

Discussion

In this national study, we elucidated three primary findings. First, the crude prevalence of EMPD in mainland China was approximately 0.04 per 100 000 population in 2016. No other studies have ever reported the prevalence of EMPD. The incidence of EMPD ranged from 0.054 patients per 100 000 person-years to 0.11 patients per 100 000 person-years in Europe based on analysis of registry database.[12, 13, 18] In the United States, the incidence was 0.07 per 100 000 person-years in Caucasian males.[1] Considering the relatively good prognosis of EMPD in those areas, with reported 5-year survival rates ranging from 50–98%, their estimated prevalence rates should be higher than our result.[11, 13, 20, 21] Within Asian area, our result was approximated to the range of rates in Taiwan of China.[22] Therefore, we could not exclude the possibility that ethnic disparity might contribute to this discrepancy, considering the fact that skin cancers are more prevalent in whites than in Asians.[23]

In our study, the prevalence of EMPD varied greatly by the geographic areas of mainland China. The Northwest China, Southcentral China and Southwest China presented significantly higher prevalence of EMPD than the rest of the areas. Based on current data, it is difficult for us to determine the exact explanation for this finding. However, this finding, to some extent, excluded the influences of socioeconomic level and medical level on the prevalence of EMPD, as the Northwest China and Southwest China are relatively underdeveloped areas in mainland China. In Europe, a clear difference in the incidence of EMPD among geographic areas were also reported, but the cause remained unclear.[13]

In addition, no other studies accessing regional difference of EMPD were available to confirm our findings, further studies are need to explain this regional discrepancy.

Secondly, patients with EMPD showed a male predominance in mainland China, with a male-to-female ratio of 1.7: 1 in our study. This was consistent with multicenter studies in Asian population, the reported male-to-female ratios were approximately 3.5:1 in Taiwan of China,[22] 3.9:1 in South Korea[24] and 2:1 in Japan.[17] By contrast, a significant female predominance was reported in studies from Western countries based on registry database, the male-to-female ratios ranged from 1:1.6 to 1:3.6.[9–13] Consistent with these gender discrepancies, Asian studies commonly identify the scrotum and penis as the most frequent sites of involvement,[24–27] rather than vulva, which is the most common site in Western populations.[10, 13] The reasons for this discrepancy remained unclear, there were two possible explanations. First, ethnic disparity might play a role, considering that Asian males were more susceptible to EMPD than Caucasian males in multiracial country.[1] Moreover, conservative attitudes among elderly Asian females that might discourage them from seeking medical treatment for lesions in the genital area was also considered an explanation in a previous study.[22]

Thirdly, EMPD were more prevalent in aged patients older than 60, with a peak prevalence at 70–79 years of age in mainland China. This was consistent with previous literatures—that is, EMPD were commonly occurred in older individuals aged 60–80.[8, 12, 14, 28] The reason for the aging of EMPD patients was unclear, a more pro-oncogenic microenvironment in aged skin might be a possible explanation.[29, 30] In addition, we found that patients with EMPD in this study were much younger than those in the United States, Europe, Japan, South Korea and Taiwan of China.[9–13, 17, 22, 24] These areas listed above had a relatively longer life expectancy than that in mainland China, which suggested that the age for EMPD patients might be closely related to the mean life expectancy in corresponding areas.[31] However, the influence of ethnic differences should also be noted, as patients with EMPD in other Asian areas including Japan, South Korea, Thailand and Taiwan of China were consistently younger than those in Western populations.[17, 22, 24, 28] Further studies are needed to clarify this point in depth.

The large, national representative sample of Chinese urban population in this study not only ensured the overall estimation of the prevalence of a rare disease like EMPD but also allowed us to explore age and gender patterns of the prevalence as well as regional differences in China. This study has several limitations. First, the basic medical insurance database lacked some detailed information, such as tumor stage and laboratory results. It limited the possibility to stratify the diagnosis in greater detail. Second, rural inhabitants and certain urban populations, such as military soldiers are not included in the UEBMI and URBMI system because they have different types of medical insurance. The exclusion of these groups could have affected the estimates.

Conclusions

This research is the first population-based study to investigate the prevalence of patients with EMPD in mainland China based on the basic medical insurance database. The prevalence of EMPD was lower in

mainland China than those in Europe and the United States. Patients with EMPD in China were younger than those in developed countries, with a significant male predominance. These findings add to our understanding of the epidemiologic characteristics of EMPD in China. At the same time, as China is a country with a population of 1.3 billion, these findings can also provide important implications for further epidemiological studies of EMPD worldwide.

Abbreviations

EMPD, Extramammary Paget's disease; UEBMI, Urban employee basic medical insurance; URBMI, Urban residence basic medical insurance; ICD, International classification of diseases; RESP, Revised European standard population; SD, Standard deviation; CI, Confidence interval

Declarations

Ethics approval and consent to participate:

The study protocol was approved by the ethical review committee of the Peking University Health Science Center (IRB. No.: IRB00001052-18012), and the informed consent requirement was waived.

Consent for publication:

Not applicable.

Availability of data and materials:

The datasets generated and/or analysed during the current study are not publicly available due to privacy, but are available from the corresponding author on reasonable request.

Competing interests:

The authors declare that they have no competing interests.

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Authors' contributions:

SY contributed to study design, data collection, data analysis, data interpretation, and manuscript writing. LX contributed to study conception, study design, data analysis, data interpretation, and manuscript writing. SW contributed to study conception, study design, administrative support, data analysis, data interpretation, and manuscript writing. JF contributed to data collection. LL contributed to data collection.

GL contributed to data collection. JW contributed to provision of study materials, and data collection. SZ contributed to study conception, study design, administrative support, data analysis, and data interpretation. ZZ contributed to study conception, data analysis, and data interpretation. PG contributed to study design, data analysis, and data interpretation. All authors read and approved the final manuscript.

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Tables

Table 1 Characteristics of the urban population from 23 provinces in China in 2016

Characteristic		Total	Male	Female
Total number (million)		425.83	221.86	203.97
Age (years)	Mean (SD)	37.68 (20.00)	37.21 (14.31)	38.20 (13.96)
Age groups, No. (% , million)	0-17	68.3 (16.04)	36.8 (16.59)	31.5 (15.44)
	18-39	167.2 (39.26)	87.7 (39.53)	79.48 (38.97)
	40-59	123.1 (28.91)	63.79 (28.75)	59.31 (29.08)
	60-69	37.89 (8.9)	19.42 (8.75)	18.47 (9.05)
	70-79	19 (4.46)	9.51 (4.29)	9.48 (4.65)
	≥80	10.35 (2.43)	4.63 (2.09)	5.72 (2.8)
Area ^a , No. (% , million)	East	171.30 (40.24)	88.68 (39.97)	82.67 (40.53)
	North	18.92 (4.44)	9.63 (4.34)	9.28 (4.55)
	Northeast	42.87 (10.07)	21.51 (9.69)	21.36 (10.47)
	Northwest	21.60 (5.07)	11.25 (5.07)	10.35 (5.07)
	Southcentral	123.60 (29.02)	66.18 (29.83)	57.41 (28.15)
	Southwest	47.50 (11.15)	24.61 (11.09)	22.89 (11.22)

Abbreviations: SD, standard deviation.

^a East area contains Jiangsu, Zhejiang, Anhui, Jiangxi, and Shandong (five provinces). North area contains Shanxi, Inner Mongolia (two provinces). Northeast contains Liaoning, Jilin, Heilongjiang (three provinces). Northwest contains Shaanxi, Qinghai, Gansu, and Xinjiang (four provinces). Southcentral

contains Henan, Hubei, Hunan, Guangdong, Guangxi and Hainan (six provinces). Southwest contains Chongqing, Guizhou and Yunnan (three provinces).

Table 2 Characteristics for patients with extramammary Paget's disease (EMPD) in 23 provinces from urban China in 2016

Characteristic	Total	Male	Female	<i>P</i> value
Number	84	53	31	
Age, years				.08
Mean (SD)	65.87 (14.21)	67.98 (12.64)	62.26 (16.13)	
Age group, No. (%)				.04
0-17	0 (0)	0 (0)	0 (0)	
18-39	4 (4.76)	1 (1.89)	3 (9.68)	
40-59	18 (21.43)	12 (22.64)	6 (19.35)	
60-69	26 (30.95)	12 (22.64)	14 (45.16)	
70-79	23 (27.38)	19 (35.85)	4 (12.90)	
≥80	13 (15.48)	9 (16.98)	4 (12.90)	
Area ^a , No. (%)				.38
East	29 (34.52)	17 (32.08)	12 (38.71)	
North	0 (0)	0 (0)	0 (0)	
Northeast	1 (1.19)	0 (0)	1 (3.23)	
Northwest	8 (9.52)	6 (11.32)	2 (6.45)	
Southcentral	31 (36.90)	22 (41.51)	9 (29.03)	
Southwest	15 (17.86)	8 (15.09)	7 (22.58)	

Abbreviation: SD, standard deviation.

^a East area contains Jiangsu, Zhejiang, Anhui, Jiangxi, and Shandong (five provinces). North area contains Shanxi, Inner Mongolia (two provinces). Northeast contains Liaoning, Jilin, Heilongjiang (three provinces). Northwest contains Shaanxi, Qinghai, Gansu, and Xinjiang (four provinces). Southcentral contains Henan, Hubei, Hunan, Guangdong, Guangxi and Hainan (six provinces). Southwest contains Chongqing, Guizhou and Yunnan (three provinces).

Table 3 Unadjusted prevalence of extramammary Paget's disease (EMPD) in urban China in 2016 (unit: /100 000 population)

	Prevalence (95% CI)
Total	0.04 (0.02-0.06)
Sex	
Male	0.05 (0.03-0.08)
Female	0.03 (0.02-0.05)
Age group	
0-17	0.02 (0.01-0.03)
18-39	0.01 (0.00-0.01)
40-59	0.04 (0.02-0.05)
60-69	0.15 (0.09-0.23)
70-79	0.28 (0.16-0.42)
≥80	0.22 (0.13-0.33)
Area	
East	0.04 (0.02-0.08)
North	0.01 (0.00-0.02)
Northeast	0.01 (0.00-0.02)
Northwest	0.07 (0.02-0.16)
Southcentral	0.06 (0.02-0.11)
Southwest	0.08 (0.03-0.16)

Abbreviation: CI, confidence interval.

Table 4 Unadjusted prevalence of extramammary Paget's disease (EMPD) in urban China in 2016, by sex (unit: /100 000 population)

	Male (95%CI)	Female (95% CI)
Age group		
0-17	0.02 (0.00-0.03)	0.02 (0.01-0.04)
18-39	0.01 (0.00-0.01)	0.01 (0.00-0.02)
40-59	0.05 (0.03-0.07)	0.03 (0.01-0.04)
60-69	0.14 (0.06-0.26)	0.16 (0.08-0.28)
70-79	0.42 (0.19-0.74)	0.10 (0.04-0.19)
≥80	0.36 (0.18-0.59)	0.15 (0.06-0.27)
Area		
East	0.05 (0.02-0.09)	0.04 (0.01-0.10)
North	0.01 (0.00-0.03)	0.01 (0.00-0.04)
Northeast	0.00 (0.00-0.02)	0.01 (0.00-0.03)
Northwest	0.09 (0.02-0.20)	0.07 (0.00-0.22)
Southcentral	0.09 (0.01-0.22)	0.03 (0.01-0.05)
Southwest	0.08 (0.02-0.18)	0.08 (0.00-0.28)

Abbreviation: CI, confidence interval.

Figures

Figure 1

Prevalence of extramammary Paget's disease (EMPD) in urban China in 2016. Note: The standardized prevalence rates are based on 2010 Chinese census data.