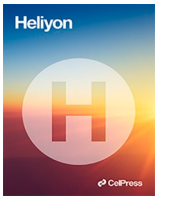
Heliyon 9 (2023) e21337

Contents lists available at ScienceDirect

Heliyon

journal homepage: www.cell.com/heliyon



Review article

A bibliometric analysis of primary Sjögren's syndrome-associated lymphoma from 1991 to 2022

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ARTICLE INFO

Keywords:
Sjögren's syndrome
Lymphoma
Bibliometric analysis
Citespace
VOSviewer

ABSTRACT

Background: Patients with primary Sjögren's syndrome (pSS) take a higher risk of developing lymphoma, which is the most frequent cause of death in pSS. Based on this situation, the number of publications focusing on pSS-associated lymphoma has been growing. Nevertheless, the extent, range, and nature of available research in this field have not been systematically summarized. This study aimed to map the literature available on pSS-associated lymphoma and identify global hotspots and trends.

Methods: Papers on pSS and lymphoma published from 1991 to 2022 were searched from the Web of Science Core Collection. Microsoft Excel, SPSS Statistics, VOSviewer, and CiteSpace software were used to analyze and visualize the quantity and citations of publications, and the global research hotspots and trends of pSS-associated lymphoma.

Results: 629 publications from 50 countries/regions and 538 institutions were included in this study. From 1991 to 2022, the cumulative publications steadily increased. The USA ranked first in the number of publications (n = 118, 18.76 %), followed by Italy (n = 94, 14.94 %) and France (n = 73, 11.61 %). Udine University (n = 29) and Salvatore De Vita (n = 39) were the most prolific affiliation and author, respectively. Claudio Vitali was the most frequently cited author (n = 335). In total, the most frequently occurring keywords were clustered into four well-defined groups. The first group of keywords pointed to the clinical assessment and treatment of pSS-associated lymphoma. The second group highlighted the pathogenesis. The third group identified the predictors and prognosis of pSS-associated Lymphoma, while the fourth group focused on interstitial lung disease and pulmonary lymphoma in patients with pSS. Currently, the hot keywords include consensus, disease activity, and pathogenesis. Ultrasonography, mucosa-associated lymphoid tissue (MALT) lymphoma, and epidemiology are the emerging research trends in pSS-associated lymphoma.

Conclusion: Research on pSS-associated lymphoma is burgeoning. Despite clinical assessment, treatment and pathogenesis, researchers also showed great interest in the predictors, prognosis, and pulmonary manifestations of pSS-associated lymphoma. Current research of pSS-associated

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https://doi.org/10.1016/j.heliyon.2023.e21337
Received 18 December 2022; Received in revised form 17 October 2023; Accepted 19 October 2023

lymphoma mainly focuses on consensus, disease activity, and pathogenesis, while the emerging research trends in pSS are pointing to ultrasonography, MALT lymphoma and epidemiology.

1. Introduction

Primary Sjögren's syndrome (pSS) is a common systemic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands resulting in sicca symptoms [1]. Although most patients with pSS presented mild disease progress, they are still at a higher risk of systemic complications such as interstitial lung disease (ILD), interstitial nephritis, and immune thrombocytopenia. What's more, the evolution into lymphoma represents one of the main causes of death in patients with pSS [2–4]. Ever since Norman Talal first described the association between SS and lymphoma more than fifty years ago, there have been countless studies carried out on this topic over the years [5]. It was reported that the risk of B-cell lymphoma is 15–20 times as high among patients with pSS as in the general population [1], and most of these lymphomas are non-Hodgkin's lymphoma (NHL) [6]. In addition, lymphomas often develop in organs such as the salivary glands in patients with active pSS, and thus are primarily mucosa-associated lymphoid tissue (MALT) lymphomas [7]. In recent years, there has been an increase in the number of studies on lymphoma in pSS. Recent research suggests that abnormal activation of B cells is the mechanism behind the occurrence of pSS, and it is also closely related to the occurrence of pSS-associated lymphoma. Zhang FC and his colleagues proposed that epithelial mesenchymal interacting protein 1 (EPSTI1) was upregulated in B cells from pSS patients, and promoted B cell proliferation and immunoglobin production, which might be implicated in the pathogenesis of pSS, even pSS-associated lymphoma [8]. Researchers are attempting to define the pathogenesis, predictors, and outcomes, so as to explore the way to prevent, predict and treat pSS-associated lymphoma [4,9,10]. However, the extent, range, and nature of available research in the field of pSS-associated lymphoma have not been systematically summarized (see Tables 1–3).

Bibliometric analysis (or bibliometrics) is an objective and reliable statistical method to quantitatively assess the academic quality of publications. It enables us to unpack the evolutionary nuances of a particular field, while illuminating the emerging areas in that field. Because of its qualities of systematic, transparent, and reproducible, this analysis has been widely used in different fields, including the medical field [11]. A bibliometric study of pSS-associated lymphoma can help us understand its current research status, progress, evolution, hotspots, and more. This information will serve as a valuable reference for future studies in this field. So far as we know, few bibliometrics studies on pSS have been published [12,13], none focused on the theme of pSS-associated lymphoma. Thus, this study aims to explore the global research hotspots and trends of pSS-associated lymphoma in the past 30 years and draw a map of scientific knowledge, so as to provide a basis for future scientific research.

2. Materials and methods

2.1. Data collection

Web of Science Core Collection (WoSCC) is one of the largest databases, which is also a more selective source for bibliometric analyses. On the other hand, WoSCC is the primary source of data for CiteSpace [14,15]. CiteSpace is optimized for handling the WoSCC data. Therefore, the data for this study were obtained from WoSCC.

We searched WoSCC on August 7, 2022. The search formula was set to [TS = (primary Sjögren's syndrome) AND TS = (Hodgkin OR non-Hodgkin OR lymphoma)]. Articles and reviews reporting lymphoma in patients with pSS were eligible for inclusion regardless of language. Early access, letters, corrections, retractions, meeting abstracts, proceedings, editorial materials, and retracted publications were excluded. As a result, 736 records were identified, of which 107 records were excluded because they didn't fulfill with the inclusion criteria. Finally, a total of 629 publications were included in this study. Fig. 1 shows the search and selection process.

Table 1
Distribution of publications in the field of pSS-associated lymphoma from different countries/regions.

Rank	Countries	Year	Centrality	Count (%)
1	USA	1992	0.15	118 (18.76)
2	Italy	1997	0.07	94 (14.94)
3	France	1991	0.12	73 (11.61)
4	Greece	1996	0.02	61 (9.70)
5	England	1993	0.12	59 (9.40)
6	Spain	1994	0.05	46 (7.31)
7	Japan	1999	0.02	43 (6.84)
8	Netherlands	1992	<0.01	42 (6.68)
9	China	1998	0.02	41 (6.52)
10	Germany	1991	0.13	36 (5.72)

2.2. Data analysis

Microsoft Office Excel 2018 and SPSS Statistics 26 (IBM, Armonk, New York, USA) were used to analyze the trend of publications in years and a p value < 0.05 was considered statistically significant. CiteSpace 6.1. R3 Advanced (Chaomei Chen, 2006) and VOSviewer 1.6.18 (Nees Jan van Eck and Ludo Waltman, 2010) were used to perform visual analysis, and create maps and clusters. CiteSpace is a visual analytic tool for visualizing landmarks, critical paths, and emerging trends in a field of research based on relevant scholarly publications. It is useful for researchers to explore the broad intellectual landscape and identify areas of interest [14,15]. CiteSpace was used to analyze the country/region, institutional distribution, author contributions, keywords burst, and co-cited references. The settings were as follows: time slicing (from 1991 to 2022), years per slice (1), selection criteria (g-index, k = 25), pruning (none). In the co-cited references map, the cluster label was selected with indexing terms and log-likelihood ratio (LLR).

VOSviewer is another bibliometric software tool for creating knowledge maps based on network data and for visualizing and exploring these maps [16]. There are three following kinds of maps in VOSviewer: network visualization, density visualization, and overlap visualization. In the network visualization map, the same color represents the same cluster, and the size of the node reflects the co-occurrence frequencies. In the density visualization map, the colours range from blue to green to yellow. The larger the number of items in the neighborhood of a point and the higher the weight of the neighboring items, the closer the color of the point to yellow. Correspondingly, the smaller the number of items in the neighborhood of a point and the lower the weights of the neighboring items, the closer the color of the point to blue. In the overlay visualization map, the color indicates the average published year. In this study, VOSviewer was used to perform keywords clustering because it was more intuitive and distinct in performing keywords clustering. The settings were as follows: counting method: full counting, minimum number of occurrences of a keyword: 5, minimum cluster size: 30.

3. Results

3.1. The trend of publications

A total of 629 papers published between January 1991 and August 7, 2022 were included in this study, involving articles and reviews. From 1991 to 2022, although the annual publications decreased in some years, the cumulative publications on pSS-associated lymphoma increased steadily year by year (Fig. 2, $p < 0.001$). In 2020, the number of annual publications reached the maximum, with a total number of 51 (8.1 %). From January to August in 2022, the number of outputs reached 18 (2.9 %), which was similar to the whole publications in 2011. All these trends indicated that research on pSS-associated lymphoma is flourishing.

3.2. Distribution of countries/regions and institutions

629 papers were published by 50 different countries/regions and 538 institutions. The United States published the most publications (n = 118, 18.76 %), followed by Italy (n = 94, 14.94 %) and France (n = 73, 11.61 %). In terms of centrality, the United States had the highest value (centrality = 0.15), followed by Germany (centrality = 0.13), and France and England (centrality = 0.12). Although Udine University published the most publications (n = 29, 4.61 %), its centrality was relatively low (centrality = 0.05). By contrast, Hospital Clinic de Barcelona (n = 15, centrality = 0.10) and Barcelona University (n = 11, centrality = 0.12) had a higher centrality. In addition, 6 of the Top 10 prolific institutions were from Italy.

3.3. Authors and co-cited authors

A total of 726 authors were involved in the publications on pSS-associated lymphoma. Salvatore De Vita was the most prolific author with the highest influence (n = 39, centrality = 0.12). When two or more authors are cited in one or more articles at the same time, they would constitute a co-citation relationship. In this situation, these authors are called co-cited authors. Among the co-cited authors, 4 had a frequency of citation over 200 times, and Claudio Vitali who was the top co-cited author (n = 335).

Table 2
Distribution of publications in the field of pSS-associated lymphoma from different institutions.

Rank	Institutions	Year	Centrality	Count (%)
1	Univ Udine (Italy)	1999	0.05	29 (4.61)
2	Univ Athens (Greece)	1998	0.05	26 (4.13)
3	Univ Pisa (Italy)	1999	0.04	20 (3.18)
4	Univ Groningen (Netherlands)	2011	0.02	18 (2.86)
5	Univ Perugia (Italy)	1997	0.03	16 (2.54)
6	Univ Aquila (Italy)	2014	0.02	15 (2.38)
7	Hosp Clin Barcelona (Italy)	2007	0.10	15 (2.38)
8	Natl & Kapodistrian Univ Athens (Greece)	2020	<0.01	15 (2.38)
9	Lund Univ (Sweden)	1999	0.06	14 (2.23)
10	Univ Barcelona (Italy)	1998	0.12	11 (1.75)

3.4. Hotspots

A total of 217 keywords were analyzed in VOSviewer 1.6.18. A keyword network map was created to establish the research topics and trends in the field of pSS-associated lymphoma. As shown in Fig. 3, every keyword is presented by labels and spheres. The higher the weight of the keyword is, the larger the label and sphere are. There are four different clusters (red, green, blue, and yellow) in the keyword network map.

The red cluster was the largest group, including 70 keywords. Classification criteria (n = 83), diagnosis (n = 46), rituximab (n = 45), and disease activity (n = 38) were the strongest keywords in this group. The green cluster included 50 keywords, and the main keywords were salivary gland (n = 113), systemic lupus erythematosus (n = 103), and disease (n = 89). The blue cluster included 49 keywords, with the main keywords: lymphoma (n = 226), primary Sjögren's syndrome (n = 171), and classification (n = 100). The yellow cluster included the rest 48 keywords. In this cluster, Sjögren's syndrome (n = 321), NHL (n = 114) and malignant lymphoma (n = 107) were the largest spheres.

3.5. Citation bursts

Strong citation bursts of keywords represent rapid changes in the number of citations for keywords, which can reflect the rise or fall of research hotspots. The period of burst is shown in red. As shown in Fig. 4, the first keyword with citation bursts first appeared in 1993, which was myoepithelial sialadenitis. Consensus, which appeared latest in 2019, had the strongest bursts (strength = 9.98). Disease activity took second place with a strength of 9.2, followed by the American College of Rheumatology (ACR) (strength = 7.2). The citation bursts of these three keywords are still continuous. Despite them, another keyword with continuous bursts of citations was pathogenesis. The continuous citation bursts mean that these topics are still hotspots at present. Therefore, consensus, disease activity, ACR, and pathogenesis are hotspots in the field of pSS-associated lymphoma at present.

3.6. The research frontier on pSS-associated lymphoma

The research frontier could be reflected by the co-citation relationship among papers. When two papers are cited by the third paper, these two papers form a co-citation relationship. Papers with citation bursts reflect the hotspots of research at some point in time. The colours of nodes and clusters represent the published year. From deep purple to light yellow, the lighter the colour is, the more proximate to the present the published year is. The light purple ring around the node represents a centrality over 0.10. The silhouette value of a cluster measures the quality of a clustering configuration. Besides, if a cluster is with a high citation and remains a high value of silhouette, it would suggest a strong reliability.

Clusters with members over 35 are shown in Fig. 5. There're 9 clusters in total, which are numbered from (#0) to (#8) by order of size. The larger the cluster is, the smaller the number is. The largest cluster (#0) labeled as MALT lymphoma had 195 members with a silhouette value of 0.894. In this cluster, the top ranked item by citation counts was Shiboski CH (2017), with citation counts of 51, followed by Theander E, 2011 (n = 41) and Fragkioudaki S, 2016 (n = 39). The second large cluster (#1) labeled as saliva had 112 members and with a silhouette value of 0.926. The third largest cluster (#2) including 107 members with a silhouette of 0.966, was labeled as pre-lymphomatous stage. Several papers with high centrality were included in this cluster, such as Anaya JM, 1996 (0.24) and Voulgarelis M, 1999 (0.22). From the colour it presented, the mean year of cluster (2#) was relatively early (1999). By the burst, the top ranked item happened to be Vitali C (2002) in Cluster (#3) labeled as hepatitis C virus. By the colours of the clusters presented, cluster (#4) ultrasonography, cluster (#0) MALT lymphoma, and cluster (5#) epidemiology are the brightest ones, which meant these three topics are supposed to be frontiers of the field of pSS-associated lymphoma.

4. Discussion

4.1. The hotspots in pSS-associated lymphoma

With Fig. 3, we could easily find four different clusters. Based on the results, the red cluster was titled as "clinical assessment and

Table 3
Distribution of author contributions in the field of pSS-associated lymphoma.

Rank	Authors	Centrality	Count (%)	Co-Cited Authors	Centrality	Count
1	Salvatore De Vita	0.12	39 (6.20)	Claudio Vitali	0.03	335
2	Xavier Mariette	0.10	29 (4.61)	Elke Theander	0.01	242
3	Athanasios G Tzioufas	0.05	26 (4.13)	Manuel Ramos-Casals	0.06	208
4	Chiara Baldini	0.08	26 (4.13)	Michael Voulgarelis	0.04	208
5	Hendrika Bootsma	0.09	25 (3.97)	Robert I Fox	0.13	193
6	Haralampos M Moutsopoulos	0.04	25 (3.97)	Stuart S Kassan	0.06	192
7	Manuel Ramos-Casals	0.05	21 (3.34)	Xavier Mariette	0.03	157
8	Luca Quartuccio	0.01	17 (2.70)	Athanasios G Tzioufas	0.17	138
9	Saviana Gandolfo	0.01	17 (2.70)	Raphaèle Seror	0.02	131
10	Clio P Mavragani	<0.01	15 (2.38)	Fotini N Skopouli	0.07	130

treatment: current status of pSS-associated lymphoma". The green cluster was named as "pathogenesis of pSS-associated lymphoma: how lymphoma occurs in patients with pSS". The blue cluster was titled as "predictors and prognosis of pSS-associated lymphoma: urgency of international multi-centre cohort establishment". At last, the yellow cluster was named as "pulmonary manifestations: interstitial lung disease and pulmonary lymphoma".

4.1.1. Red cluster: clinical assessment and treatment: current status of pSS-associated lymphoma

This cluster presented the current status of pSS-associated lymphoma involving clinical diagnosis and treatment. Common symptoms, clinical examinations, classification criteria, and disease activity could also be found in this cluster.

Sicca symptoms, such as dry eyes and dry mouth are the most typical symptoms of pSS. When patients complain of these symptoms, it suggests that they may have pSS. When pSS is suspected, examinations such as ultrasonography, sialography, and salivary gland biopsy are helpful. Ultrasonography and sialography can be used to assess the construction and function of salivary glands. Moreover, ultrasonography is helpful in the diagnosis of lymphoma. Biopsy for pSS and pSS-associated lymphoma includes minor salivary gland biopsy (MSGB), parotid gland biopsy, and lymph node biopsy. Sialography has played an important role in the diagnosis of pSS in the past, but its current application is very limited. Due to the growing number of inspection techniques and associated risks, it was excluded from the 2016 ACR and European League Against Rheumatism (EULAR) classification criteria [17].

MSGB is a minimally invasive and safe surgical procedure, which is considered as one of the main items for the diagnosis of pSS [18]. By taking this examination, the focus score (FS) and the number of germinal centers (GCs) could be calculated. FS and GCs were proved to be associated with the incidence of lymphoma in patients with pSS. The presence of GCs strongly increased the risk of lymphoma development. FS is the total number of foci per 4 mm^2 of salivary gland tissue and a focus is defined as an aggregate of ≥ 50 lymphocytes. The use of FS is not only for diagnosis but also for prognosis, especially for lymphoma developing. FS on pSS diagnosis was proven an independent predictor for MALT lymphoma in recent research [19]. FS ≥ 3 or ≥ 4, was respectively reported as a possible predictive marker for early diagnosis of pSS-associated lymphomas [20,21]. The authors concluded that the higher the FS is, the higher the risk of pSS developing lymphoma is [20,21]. Because of the predictive value, some researchers suggest that MSGB should be a regular examination for patients with pSS, no matter whether they are in the absence of anti-SSA antibody or not. However, some researchers held a different opinion. A Korean prospective cohort study enrolling 161 pSS patients with FS ≥ 1 and another 24 with FS < 1, found that there were no significant differences between the two groups in the risk of lymphoma. Therefore, researchers suggested that MSGB could be omitted for serologically and clinically established pSS patients [22]. It was reported that parotid gland biopsy was necessary for the detection of MALT lymphoma in patients with pSS. When pSS patients repeat frequent symptoms of parotid enlargement, parotid gland biopsy should be considered [23]. Moreover, the sensitivity and specificity of parotid gland biopsy and MSGB were comparable [24]. Thus, some researchers suggested replacing MSGB with parotid gland biopsy.

It's worth mentioned that ultrasound-guided core needle biopsy (CNB) is another advanced examination for pSS-associated lymphoma. It is recommended for pSS patients with salivary gland enlargement. Compared with open surgical biopsy, US-guided CNB is

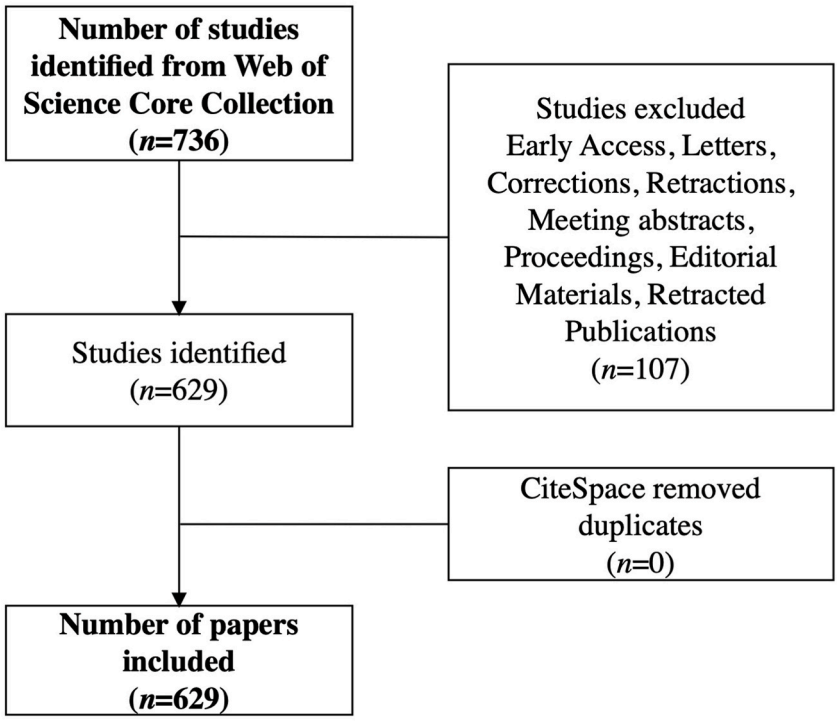

Fig. 1. Flowchart of literature search and selection.

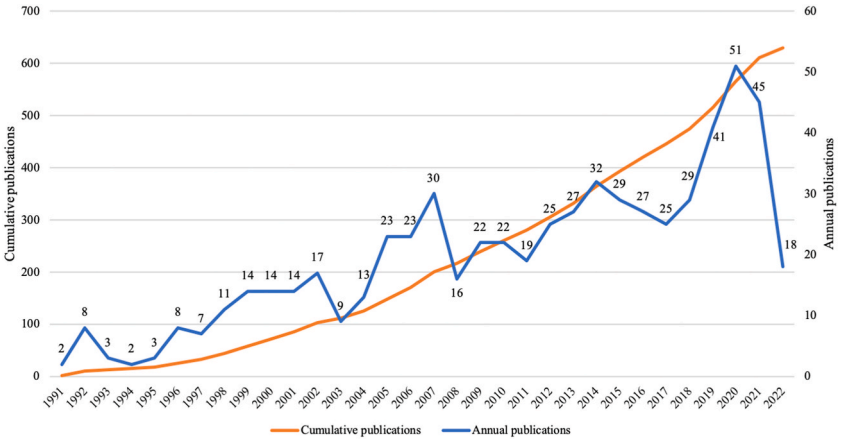

Fig. 2. Trends of publications in the field of pSS-associated lymphoma from 1991 to 2022.

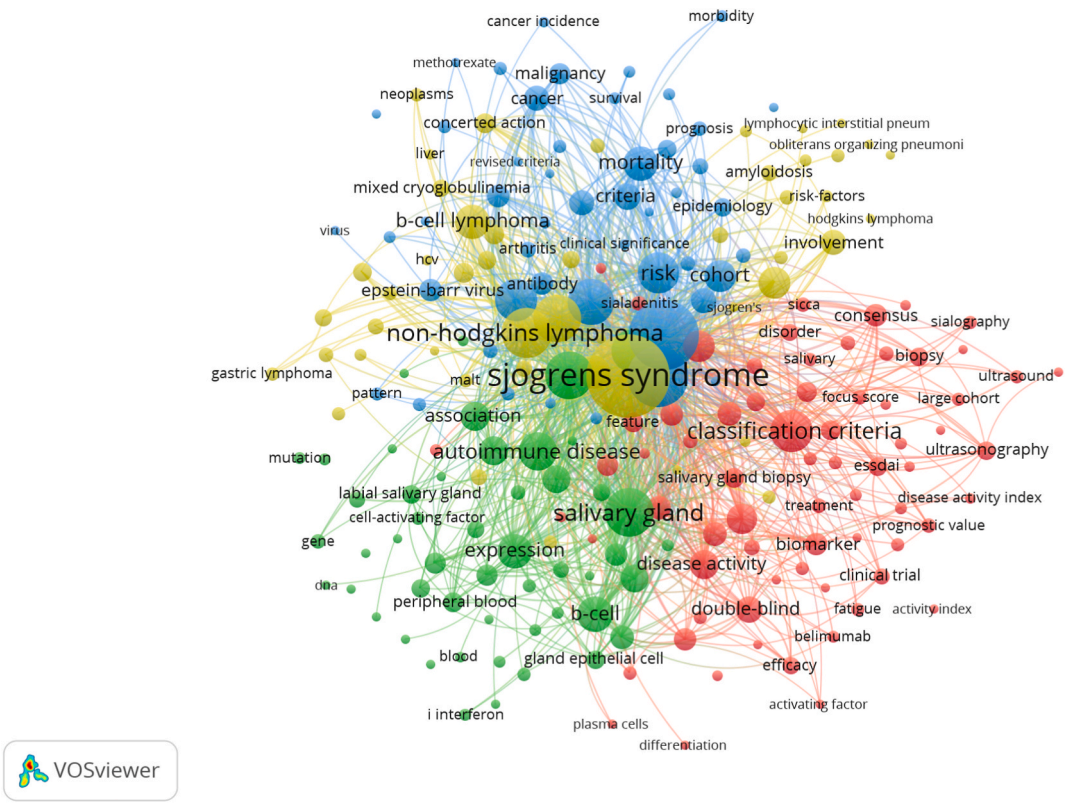

Fig. 3. Network map of keywords related to pSS-associated lymphoma.

safer and easier to be accepted by patients, and is equally effective. Unfortunately, it has not yet been widely adopted, and the sample size in current research is relatively small [25]. Nonetheless, it's undeniable that its potential is worth further exploration in the future.

Although biopsy showed a high value in both diagnosis and prognosis, it is still an invasive procedure. To reduce patients' suffering from invasive procedure, salivary gland ultrasonography (SGUS) has gradually become a research hotspot recently. SGUS is a harmless, convenient, and performant way to examine the structure of salivary gland. There are four major scoring tools designed for SGUS in the assessment of pSS: the Salaffi score, Jousse-Joulin score, Hocevar score, and Milic score. These measurements were all corroborated to show a good diagnostic performance for pSS [26]. According to previous research, there was a significant correlation between SGUS score and FS, which suggested that SGUS may replace MSGB in the future [27,28]. Recently, the value of SGUS has been highly evaluated and suggested to use in daily practice for patients with pSS. In addition, it was mentioned that SGUS might be a possible predictor for pSS patients developing lymphoma, which needs to be proved in prospective studies [29]. Therefore, more

Keywords	Year	Strength	Begin	End	1991-2022
myoepithelial siadenitis	1991	4.49	**1993**	2002	
criteria	1991	5.06	**1996**	2002	
helicobacter pylori	1991	4.34	**1996**	2002	
gastric lymphoma	1991	4.01	**1996**	2002	
hepatitis c virus	1991	6.15	**1997**	2008	
concerted action	1991	5.36	**2001**	2008	
malignant lymphoma	1991	3.90	**2001**	2004	
primary biliary cirrhosis	1991	3.68	**2001**	2003	
rheumatoid arthritis	1991	3.58	**2004**	2007	
therapy	1991	5.50	**2005**	2007	
non-hodgkins lymphoma	1991	5.73	**2006**	2008	
cohort	1991	3.79	**2008**	2011	
rituximab treatment	1991	5.37	**2010**	2018	
mortality	1991	4.29	**2010**	2013	
gland epithelial cell	1991	3.86	**2011**	2015	
lymphoma development	1991	6.25	**2012**	2017	
classification criteria	1991	5.61	**2014**	2015	
pathogenesis	1991	5.02	**2014**	2022	
rheumatoid factor	1991	4.57	**2015**	2019	
disease activity	1991	9.20	**2017**	2022	
american college of rheumatology	1991	7.20	**2018**	2022	
consensus	1991	9.98	**2019**	2022	

Fig. 4. Top 22 keywords with the strongest citation bursts.

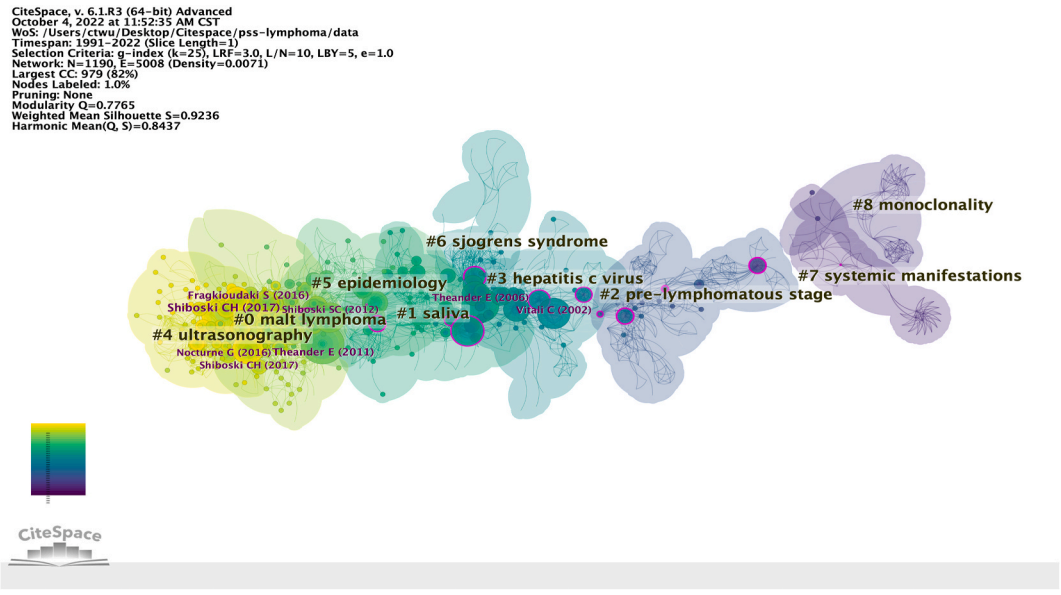

Fig. 5. The visualization of co-cited references in pSS-associated lymphoma.

efforts are needed to assess the value of SGUS in pSS and pSS-associated lymphoma.

Since 1975, more than seven criteria from different countries have been developed for the diagnosis of pSS. Of which, the 2002 American-European Consensus Group criteria and the 2016 ACR/EULAR classification criteria are the most widely used classification criteria for pSS [30]. With further research, updated diagnostic criteria may be generated in the future.

There are two commonly used international indexes for the assessment of disease activity in pSS, including the EULAR Sjogren's Syndrome Patient Reported Index (ESSPRI) and EULAR Sjögren's Syndrome Disease Activity Index (ESSDAI). ESSPRI is a patient-administered questionnaire involving three visual-analogue scales to assess dryness, fatigue, and pain [31]. ESSDAI is an important tool to assess systemic complications of pSS in 12 domains but is often used in clinical trials [32–34]. Whether ESSDAI can be a predictor for lymphoma in pSS patients or not, is also a controversial topic. In 2016, a multicenter case-control survey including 101

French or British pSS patients with lymphoma, highlighted that the ESSDAI was as an independent predictor for lymphoma in pSS [35]. Five years later, a cohort study enrolling 878 Greek pSS patients, mentioned that biologic and glandular ESSDAI domains could reflect the lymphomagenesis course towards MALT lymphomas [18]. Another cohort study on Spanish pSS patients with high ESSDAI showed that this subset of patients had a higher mortality rate of 20 %, which was mainly caused by lymphomas [36]. On the other hand, an Italian cohort study including 255 pSS patients found that ESSDAI was not an ideal tool for the assessment of lymphoma, because it showed no significant differences between pSS patients evolving into lymphoma and those who did not [37]. Due to the inconsistent findings, some researchers suggest designing a targeted tool for pSS patients to assess their risk of developing lymphomas.

There is no definite consensus exists on the treatment of pSS-associated lymphoma. Treatments are chosen based on the pathologic result as lymphomas in the general population. In this cluster, rituximab (RTX) presents a high weight in 70 keywords, which means that RTX is a certain hotspot in the field of pSS-associated lymphoma. RTX is an anti-CD20 monoclonal antibody, which is often used in treating lymphomas (mostly NHL), leukemias, and autoimmune diseases. It has been reported that RTX was an effective treatment for pSS-associated MALT lymphoma, however, only in case reports [38–40]. There's still no high-quality evidence for RTX in treating pSS-associated lymphoma.

4.1.2. Green cluster: pathogenesis of pSS-associated lymphoma: how lymphoma occurs in patients with pSS

Pathogenesis has always been a hotspot in the field of pSS, particularly pSS-associated lymphoma. In this cluster, the frontiers of pathogenesis in the field of pSS-associated lymphoma included nuclear factor kappa-B (NF-κB), b-cell activating factor family (BAFF), interferons (IFNs), and GCs.

Evidence suggests that activation of NF-κB pathway is a critical step in the pathogenesis of both pSS and B-cell NHL. Two different cohort studies including English, French and Greek pSS patients confirmed that tumor necrosis factor alpha-induced protein 3 (TNFAIP3) gene was associated with pSS-associated lymphoma [41,42]. TNFAIP3 gene encodes the A20 protein, which is an important negative feedback regulator of the NF-κB pathway. The variants of TNFAIP3 could lead to the abnormal control of NF-κB activation in B-cells continuously stimulated by autoimmunity, which enhances the risk of lymphoma. While in other nations, studies on TNFAIP3 gene on pSS-associated lymphoma remain a blank.

BAFF also plays a critical role in pSS-associated lymphoma. Nocturne, G. and Mariette, X. suggest that BAFF secretion could stimulate B-cells by a positive loop of activation, which can promote the development of lymphoma in patients with pSS [4]. The role of IFNs in the pathogenesis of pSS has been proved. Nezos, A. and his colleagues pointed out that type I-IFN (IFN-α) and type II-IFN (IFN-γ) could be associated with the development of lymphoma in pSS. In their work, the high level of IFN-γ and the low level of IFNα in minor salivary gland (MSG) tissues were related to lymphoma. Thus, the IFN-γ/α mRNA ratio in MSG was proposed to be a biomarker for lymphoma in pSS [43]. Additionally, IFN-α is known to be involved in B-cell differentiation by its upregulation of BAFF [44,45]. Ectopic GCs and GC-like structures in MSG tissues have been proved to be predictors for lymphoma in pSS patients in several studies [45,46]. It was reported that pSS patients who present ectopic GC-like structures in MSG tissues presenting a 7.8-fold increased risk of developing lymphoma [46]. On the other hand, Haacke EA and his colleagues found that GCs in MSGB is not a predictor for pSS-associated parotid MALT lymphomas by comparing 11 labial gland biopsies from patients with pSS that were taken prior to parotid MALT lymphoma development and biopsies of 22 matched pSS controls who did not develop lymphoma [47].

4.1.3. Blue cluster: predictors and prognosis of pSS-associated lymphoma: urgency of international multi-centre cohort establishment

Despite clinical diagnosis and pathogenesis, researchers and clinical doctors showed more interest in clinical predictors and prognosis of pSS-associated lymphoma. Hence, cohorts of pSS patients have been built up in different nations to explore these.

A series of clinical predictors of pSS-associated lymphoma have been discovered [48]. Salivary gland enlargement (SGE) especially parotid swelling [35], lymphadenopathy [49], Raynaud phenomenon [48], peripheral neuropathy [50], and purpura [51] are signs and symptoms that have been mentioned as clinical predictors of lymphoma in patients with pSS. Meanwhile, rheumatoid factor [35,49], cryoglobulinemia [51], TNFAIP3 gene [41], and hypocomplementemia (low level of C3 or C4 complement) [50,51] are serological predictors of lymphoma. As mentioned above, FS [22–24], GC-like structures [21], and IFN-γ/α mRNA ratio [43] in MSGB are also potential predictors of developing lymphoma in pSS. Interestingly, among these predictors, SGE, peripheral neuropathy, purpura, cryoglobulinemia, and hypocomplementemia are also included in the assessment subjects of ESSDAI. Thus, some scholars suggested that ESSDAI could be used as a predictive tool for lymphoma in pSS [35]. Besides, some studies showed that by different onset ages of pSS, patients had different risks of developing lymphoma: the young onset pSS patients with cryoglobulinemia, lymphadenopathy, and SGE took higher risks, while the late onset ones with male gender had higher risks [52].

Another challenge for researchers is how to identify potential prognostic factors for pSS-associated lymphoma. Compared with the general population, lymphoma slightly increased the mortality rate of pSS patients [53]. A retrospective study concluded that nodal marginal zone lymphoma (NMZL) in pSS patients showed poor outcomes [54]. Another study showed that both ESSDAI and international prognostic index (IPI) score presented a significant prognostic role in survival outcomes of SS-associated lymphoma [55].

In the last 30 years, several independent cohorts of pSS patients have been set up in different countries to explore the clue to poor prognosis. Despite a series of research have been published, many questions related to predictors and prognosis of pSS-associated Lymphoma remain unresolved. Patients with pSS have a high risk of lymphoma and other malignancies, thus it is necessary to establish an international multi-centre cohort to study this. Cooperation across borders would certainly have an overall positive effect on research in the field of pSS and its complications.

4.1.4. Yellow cluster: pulmonary manifestations: interstitial lung disease and pulmonary lymphoma

To our best knowledge, pSS is associated with various pulmonary manifestations, especially different kinds of ILD, mostly

lymphocytic interstitial pneumonia (LIP). It is reported that pulmonary lymphoma was relatively rare, for it represents about 4 % of all extra-nodal lymphomas. But when it comes to patients with pSS, the prevalence seems to be higher and is estimated to be 1–2%. Pulmonary lymphoma in pSS patients is usually a low-grade extra-nodal marginal B-cell lymphoma of the MALT type [53]. Because of the similarities in manifestations but differences in treatments between ILD and pulmonary lymphoma, it is necessary to distinct the two diseases as early as possible. Usually, clinician can make a preliminary diagnosis based on the radiologic features for those with significant lesions. To make an accurate diagnosis, a lung biopsy and a pathological examination should be performed. Immunohistochemical and molecular techniques were also very useful diagnostic methods. However, the early diagnosis technology still needs to be improved. Additionally, it is noteworthy that about 5 % of LIP patients progress to lymphoma [56]. For the relationship between ILD and pulmonary lymphoma in patients with pSS, there is no conclusion. Whether pSS patients with ILD have a higher risk of lymphoma or not remains unclear. More studies are needed to further explore this.

4.2. Comparison between bibliometric analysis on (p)SS and pSS-associated lymphoma

As mentioned earlier, bibliometric studies on (p)SS have been done [12,13]. When comparing the previous studies to our study, a noticeable disparity is observed. In the research solely focused on (p)SS, China has a rich publication output and ranks second. However, in this study, its position is relatively lower, occupying the ninth position. We speculate that this discrepancy may be attributed to China's predominant focus on researching the pathogenesis of (p)SS, treatments including traditional Chinese medicine and western medicine, and ILD. In contrast, there is less research from China on pSS-associated lymphoma.

4.3. Future of pSS-associated lymphoma

Lymphoma, as one of the most severe complications of pSS, it is crucial to understand its mechanisms, identify high-risk populations, and achieve early diagnosis. In the future, if we can prevent the occurrence of pSS-associated lymphoma based on clarifying its mechanisms, many patients will undoubtedly benefit. At the same time, strengthening connections between countries and establishing international multi-center cohorts to summarize population charateristics, identify risk factors, and develop predictive models will be a trend.

5. Strengths and limitations

This study firstly mapped the literature available on pSS-associated lymphoma and identify global hotspots and trends in this field. However, our study only involved published articles on pSS-associated lymphoma in WoSCC, those from other databases were not included, which may lead to a bias. In addition, the citation of 2022 is lower than previous years. Furthermore, the quantity of publications may be affected by countries with smaller populations or lower GDPs, resulting in fewer publication contributions. These could all be potential flaws in the bibliometric study.

6. Conclusions

This is the first bibliometric analysis drawing a map of scientific knowledge on pSS-associated lymphoma in the past 30 years. We systemically summarized the global hotspots and trends in this field. The results showed that research on pSS-associated lymphoma is burgeoning. Despite clinical assessment, treatment and pathogenesis, researchers also showed great interest in the predictors, prognosis, and pulmonary manifestations of pSS-associated lymphoma. Current research of pSS-associated lymphoma mainly focuses on consensus, disease activity, and pathogenesis, while the emerging research trends in pSS are pointing to ultrasonography, MALT lymphoma and epidemiology. It is necessary to conduct international cooperative study to improve the survival and prognosis of pSS-associated lymphoma.

Data availability statement

Data sharing is not applicable to this article as no new data were created in this study. These datasets were derived from the following public domain resources: http://www.webofscience.com/wos/woscc/advanced-search.

CRediT authorship contribution statement

Tzuhua Wu: Conceptualization, Data curation, Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Shangdian Li:** Data curation, Methodology, Writing – original draft. **Jiaqi Chen:** Data curation, Methodology, Writing – original draft. **Jiahe Liao:** Data curation, Methodology. **Ziwei Huang:** Data curation, Methodology. **Jianying Yang:** Data curation. **Yan Zhang:** Data curation. **Qian He:** Data curation. **Xinbo Yu:** Data curation. **Weijiang Song:** Data curation. **Jing Luo:** Conceptualization, Data curation, Methodology, Writing – review & editing. **Qingwen Tao:** Conceptualization, Methodology, Writing – review & editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to

influence the work reported in this paper.

Acknowledgements

Author would like to thank CiteSpace and VOSviewer software developers. This work was supported by National High Level Hospital Clinical Research Funding (2022-NHLHCRF-LX-02-0103), and Elite Medical Professionals project of the China-Japan Friendship Hospital (ZRJY2021-QM14), the Capital's Funds for Health Improvement and Research (2020-4-40610).

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