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Title

Paraneoplastic Syndromes Associated with Classic Hodgkin Lymphoma; A systematic Literature Review

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Introduction

Cancer signs that are unrelated to direct tumor infiltration or metastasis are defined as Paraneoplastic Syndromes (PNS). PNS can have a wide range of effects on organ systems, including the dermatologic, neurologic, hematologic, endocrine, renal, rheumatologic, and hepatic systems, Although a specific diagnostic criterion was created to aid in the diagnosis of PNS, available literature on PNS is lacking and mostly made up of case reports and small case series. This systematic review aims to gather all empirical evidence related to PNS and summarize it in order to better understand these perplexing disorders.

Methodolgy

Literature searches were performed in the following databases: PubMed, Web of Science, SCOPUS, MEDLINE and CINAHL. Studies published from January 2000 until October 2021 using the following MESH Terms “paraneoplastic syndrome” AND “Hodgkin disease” were included. References of previous reviews on PNS were also screened to ensure the inclusion of all relevant literature. Data extraction and eligibility screening were performed and analyzed using SPSS Statistics.

Results

Our literature search yielded 1030 unique citations; we ended up with 836 citations to be screened. After screening, 123 reports were included for analysis. Of these, 115 were case reports and eight were case series with a total number of patients of 128. Adult age group (>18 years) had a statistically significant association with higher prevalence of PNS as compared to pediatrics age group (P=0.001) with the majority of patients being males. The most frequently reported ethnicity was Caucasian. The most commonly reported cHL histologic subtype was NS subtype (66.4%) and the most frequent disease stage at diagnosis was stage II (33.6%), 28.1% had extra nodal involvement at presentation. Central nervous system manifestations were the most frequent clinical presentation (25.8%) . In 33.6% of patients, the lymphoma diagnosis preceded the PNS diagnosis. While the PNS diagnosis preceded the lymphoma diagnosis in only 16.4% of patients. The presence of PNS antibodies was reported in 27.3% of patients with the NS subtype having a statistically significant association with higher incidence of PNS antibodies (P=0.004). Majority of patients (47.6%) were treated with a combination therapy targeting both the PNS and the lymphoma. The complete remission rate of the lymphoma was 77.3%. The complete resolution rate of the PNS was 54.7%. Relapse of lymphoma was reported in 13 patients and recurrence of the PNS upon relapse was reported in 10 out of 13 patients.

Conclusion

In this systematic review of published reports, we tried to draw a collective integrated appraisal of a rare co-occurrence that is rarely addressed in clinical trials. The following suggestions have been made: (1) An international collaborative effort is needed to build a large and comprehensive database that can help in developing standard guidelines and recommendations to help in the understanding of PNS and aid in the diagnosis and management of patients with PNS. (2) Bio banking is another critical component that may help discover new antibodies or to study the significance of omics (proteomics, metabolomics, etc.) in these patients. (3) Finally, a high degree of suspicion and awareness among healthcare providers is needed to capture these cases and subsequently provide the appropriate measures.