

Sinonasal extramedullary plasmacytoma: a systematic review of 175 patients

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Background: This study reviews the published literature related to extramedullary sinonasal plasmacytomas. Clinical presentation, demographics, treatment, and outcomes of this uncommon disease are reported.

Methods: A systematic review of studies for sinonasal plasmacytomas from 1950 to 2012 was conducted. A PubMed database search, both for articles related to this condition along with bibliographies of those selected articles, was performed. Articles were examined for patient data that reported disease outcome.

Results: Sixty-seven journal articles were included in this analysis, comprising a total of 175 cases. Radiotherapy was the most common treatment modality, used in 89 cases, followed by a combination of surgery and radiotherapy, and surgery alone. A total of 71.8% of patients were alive after a median follow-up of 39 months, independent of treatment modality. A combination of radiotherapy and chemotherapy was rarely used but had the best treatment outcome, with 88.9% of patients (8/9 patients) alive. Of the 3 most common treatment modalities, a combination of radiotherapy and surgery had the most favorable outcomes. Sixteen

patients (9.1%) converted to multiple myeloma, with the majority of these patients (75.0%) receiving radiotherapy alone as their treatment modality.

Conclusion: This review contains the largest pool of sinonasal plasmacytoma patients to date and suggests aggressive radiotherapy is the most common treatment modality for this condition. Of the 3 most common treatment modalities, a combination of surgery and radiotherapy was shown to have the best survival outcomes. © 2013 ARS-AAOA, LLC.

Key Words:

sinonasal plasmacytoma; extramedullary plasmacytoma; nasal tumor; sinus tumor; plasmacytoma; radiotherapy; chemotherapy; surgical management

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Plasmacytomas are malignant neoplasms of monoclonal B-cell proliferation, consisting of 3 types: multiple myeloma (MM), solitary bone plasmacytoma (SBP), and extramedullary plasmacytoma (EMP).¹⁻³ EMP is a rare entity, accounting for only 3% of all plasmacytoma cases. As the name suggests, EMP develops outside the skeletal system as a soft tissue neoplasm.⁴ The rarity of EMP is

demonstrated by its low worldwide incidence, at 0.04 to 3 cases per 100,000. It accounts for 1% of all head and neck tumors and 4% of non-epithelial neoplasms of the sinonasal tract.^{1,4,5} EMPs are 4 times more common in men and tend to affect the elderly, with the highest incidence of new cases reported in the 6th to 8th decade of life.¹⁻⁴ Although a rare tumor overall, 78% to 80% of cases occur within the head and neck region, with approximately 80% of those occurring within the sinonasal cavity.^{1,5,6}

Various therapeutic modalities for the treatment of EMP have been suggested. However, the course of treatment has not been standardized due to rarity of the tumor, its frequent occurrence in the elderly population with common comorbidities, and the suggestion that EMP be treated as an aggressive malignancy.^{8,9} Treatment is often personalized to the individual patient in order to best preserve the intricate anatomy from where these tumors typically arise. EMPs are radiosensitive, and radiotherapy is often used as a first-line treatment to avoid disfiguring results and potentially eliminate the need for further invasive surgical

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procedures. In small, localized cases, however, complete surgical excision is appropriate. The role of chemotherapy in the treatment of EMP is controversial. Newer studies have shown that adjuvant chemotherapy has not improved survival rates, reduced relapses, or slowed the progression to MM.^{8,9}

The development of MM is an uncommon sequelae of EMP. It is believed that only 15% to 20% of EMP cases progress to MM or regional disease.⁵ The development of MM is far more common in SBP, with conversion rates estimated between 48% and 85% of SBP cases.¹ Factors that promote the conversion of EMP or SBP into MM include expression of the lambda chain within tumors, presence of greater than 10% of plasma cells in bone marrow, and involvement of multiple sites.¹

Sinonasal plasmacytomas are rare tumors. Consequently, there is little published evidence regarding the epidemiology, optimal management, and long-term prognosis for these tumors. Due to their rarity, it is challenging to undertake a randomized controlled trial to determine the optimal management of sinonasal EMPs. Therefore, this article undertakes a systematic review of 175 cases of EMPs of the sinonasal tract, representing the largest single collection of sinonasal EMPs to date. We aimed to analyze the demographics, presentation, treatment modalities, prognosis, and potential sequelae of this rare tumor.

Materials and methods

Search strategy

A systematic review, using the PubMed database, was conducted for all cases of sinonasal plasmacytoma from 1950 to 2012. Search criteria were input as “sinonasal plasmacytomas,” “sinus plasmacytomas,” and “nasal plasmacytomas”; 512 initial articles were obtained. Articles were then filtered to exclude nonhuman and non-English language research. Abstracts were first reviewed to search for articles that discussed cases of plasmacytomas occurring in the sinonasal tract, and full-text articles were subsequently selected and reviewed for extraction of data. References of the included studies were examined for additional cases. This review qualified as “nonhuman subject research,” and did not require approval by the institutional review board.

Selection criteria and data extraction

All English, human studies that reported individual data for sinonasal plasmacytomas were included if they reported diagnosis, treatment, follow-up, and outcome. Nonhuman, radiologic, cadaveric, anatomical, and histological studies were excluded, as were sources with insufficient or unextractable data. Articles with unobtainable full text were also excluded. Outcome measures extracted included: demographic data, tumor location, presenting symptoms, radiographic imaging, primary treatment modality, adjuvant treatment, recurrence, metastasis, development of multiple myeloma, follow-up, secondary treatment for recurrence

or metastasis, and overall survival. This analysis used Microsoft Excel (Microsoft Corp., Redmond, WA) for data aggregation and analysis.

Results

The initial PubMed searches using the keywords “sinonasal plasmacytoma,” “sinus plasmacytoma,” and “nasal plasmacytoma” yielded 512 studies. Studies that were in a non-English language or pertaining to animal research were excluded (218). Next, duplicate articles were eliminated (35). For the articles remaining, the abstracts were reviewed for relevance. Those abstracts that did not pertain to the subject matter, discussed a different diagnosis, or focused on a different anatomical location were eliminated (129). Studies were then examined for individual patient data, and if this data could not be extracted, the article was excluded (29). Articles with individual patient data but focused solely on histologic, radiologic, or pathologic findings were excluded (15). Finally, articles in which the full-text could not be obtained were eliminated (19). Of the remaining articles, the bibliographies were examined for additional relevant articles that met the criteria outlined above (Fig. 1). Sixty-seven studies comprising a total of 175 patients were left for analysis. All 67 studies included individual patient data that was extractable and fit the minimum criteria for inclusion. The 67 studies are detailed in Table 1.^{2,5,8,10-73}

Demographics

Patient demographics for the 175 patients included in this review are summarized in Table 2. The mean age of patients was 55 years, ranging from 5 to 79 years. Males comprised 69.4% of cases, whereas females comprised 30.6% of cases. The mean and median follow-up for patients was 60.9 and 39 months, respectively. Information about tumor size, stage, and histological grade was rarely reported and thus was not included in this analysis. The most common anatomic site of the primary tumor was the nasal cavity and septum (32.5% of cases) and the most common presenting symptom was nasal obstruction (29.8% of cases). Further demographic information is summarized in Table 2.

Treatment modalities and outcome

Treatment modality was reported in all articles used in this review and included radiotherapy, surgery, chemotherapy, or various combinations of these modalities. The most common treatment modality was radiotherapy alone, used in 89 cases (50.9%). A combination of surgery and radiotherapy was the next most common treatment type, used in 38 cases (21.7%). Surgery alone was the treatment of choice in 25 cases (14.3%), and chemotherapy alone was used to treat 3 cases (1.7%). No patients received a combination of surgery and chemotherapy as treatment, and 5 patients (2.9%) did not receive any treatment. Further information regarding treatment modality is summarized in Table 3.

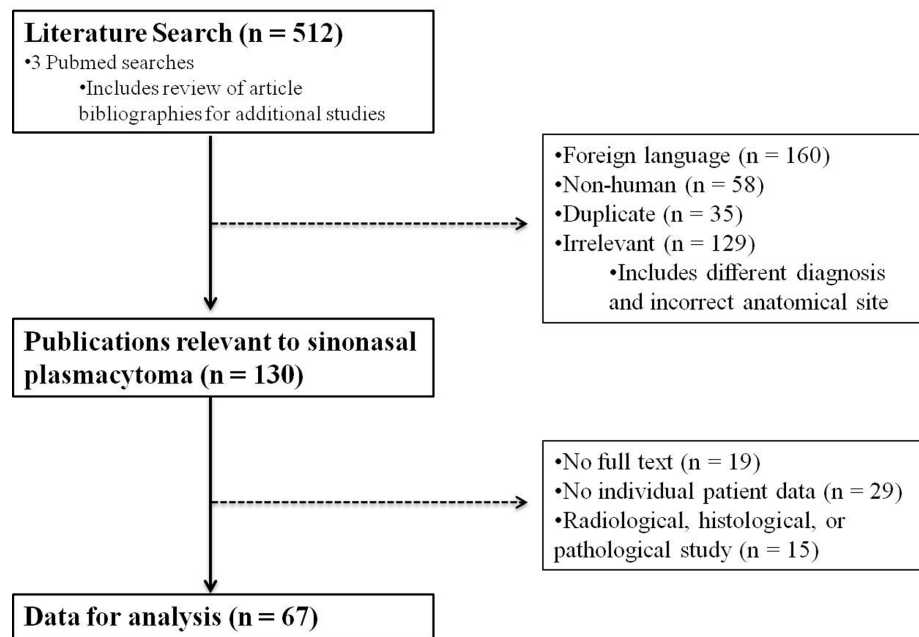


FIGURE 1. Selection process during primary searches. n = number of studies.

Patient outcomes in this study were classified as either alive with no evidence of disease (ANED), alive with disease (AWD), died of disease (DOD), and died not of disease (D). Overall outcome was generally favorable, as 100 patients (57.5%) were alive with no evidence of disease and 25 patients (14.3%) were alive with disease after a median follow-up of 39 months. A total of 48 patients (27.4%) died, with 24 of these patients (13.8%) dying from disease.

Table 4 details patient outcomes by treatment modality. Patients receiving a combination of radiotherapy and chemotherapy had the most favorable outcome, with 88.9% (8/9 patients) being classified as ANED or AWD. However, of the 3 most common treatment modalities (radiotherapy alone, surgery and radiotherapy, and surgery alone), patients receiving a combination of surgery and radiotherapy had the most favorable outcome, with 81.6% (31/38 patients) being classified as ANED or AWD. The outcome for patients receiving radiotherapy alone was ANED or AWD for 69.7% (62/89 patients), and for patients receiving surgery alone was 68% (17/25 patients).

Thirty-one articles contained 104 cases from the year 1990 and earlier and 36 articles contained 71 cases from 1991 onward. In the cases reported before and including the year 1990, the most common treatment modality was radiotherapy as a single modality (67 cases, 64.4%), followed by combination radiotherapy and surgery with 22 cases (21.2%), 7 cases of surgery alone (6.7%), 5 cases of combination radiotherapy and chemotherapy (4.8%), 1 case of chemotherapy alone (1.0%), 1 case of combination radiotherapy, surgery, and chemotherapy, and 1 case that received no treatment. The most common treatment modality from 1991 onwards was also single-modality radiotherapy, accounting for 25 cases (35.2%). Seventeen patients

(23.9%) were treated with surgery alone, 17 with combination radiotherapy and surgery (23.9%), 6 with combination radiotherapy, surgery, and chemotherapy (8.5%), 4 with a combination of radiotherapy and chemotherapy (5.6%), and 2 with chemotherapy alone (2.8%).

In the 22 articles that reported recurrence or development of multiple myeloma, 28 patients (16.8%) developed recurrence of disease. Sixteen patients (9.1%) developed multiple myeloma, of which the majority of these patients (12 patients, 75%) received radiotherapy as their treatment modality.

Discussion

EMP are rare entities, accounting for only 3% of plasma cell neoplasms, and 1% of head and neck tumors.^{4,10} The most common location for EMP is the head and neck region, with 80% of cases occurring in the rich lymphatic tissue of the upper respiratory tract, including the nose and paranasal sinuses.¹⁰ These tumors tend to present insidiously, with nonspecific, obstructive symptoms, delaying timely diagnosis of the tumor.¹¹ This study represents the first systematic review of sinonasal EMPs, providing meaningful insight into this rare tumor.

Previous publications have shown that sinonasal EMPs commonly affect males in a 3:1 ratio and present between the 4th and 7th decade of life.^{10,72,74} Our data also showed a higher occurrence in men, with a male to female ratio of 2.3:1. We found that the mean age of diagnosis was 55 years, with a range of 5 years to 79 years, consistent with previously published data. Sinonasal EMPs most often present as a space-occupying lesion, with nasal obstruction being the first sign of the malignancy. We did find

TABLE 1. Included studies

Author	Year	Study type
Abemayor et al. ¹²	1988	Case series
Ampil et al. ¹³	2002	Case report
Araújo Rde et al. ¹⁴	2008	Case series
Attanasio et al. ¹⁵	2006	Case report
Baek et al. ¹⁶	2005	Case report
Batsakis et al. ¹⁷	1964	Case report
Booth et al. ¹⁸	1973	Case report
Buchanan et al. ¹⁹	1972	Case series
Cakir et al. ²⁰	2003	Case report
Chaudhuri et al. ²¹	1988	Case report
Cumbo et al. ²²	1988	Case report
de Gast et al. ²³	1976	Case report
Dempewolf and Lee ⁷¹	2008	Case report
Erkal et al. ⁸	2006	Case report
Exarchakos et al. ²⁴	2003	Case report
Ferlito et al. ²⁵	1982	Case report
Fu and Perzin ²⁶	1978	Retrospective review
Ghildiyal et al. ²⁷	1990	Case report
Greenberg et al. ²⁸	1987	Clinical Investigation
Gross et al. ²⁹	2004	Case report
Grover et al. ⁵	2006	Case report
Hasegawa et al. ³⁰	1987	Case series
Hazarika et al. ¹⁰	2011	Case series
Helmus ³¹	1964	Case report
Humphrey et al. ³²	1983	Case report
Jay ³³	1955	Case report
Jeon et al. ³⁴	2003	Case series
Kanimiturk ³⁵	1978	Case report
Kayrouz et al. ³⁶	1983	Retrospective review
Kelly and Lloyd ³⁷	1991	Case report
Ko et al. ³⁸	2009	Case report
Kondo et al. ³⁹	1986	Case series
Kondo et al. ⁴⁰	1995	Case report
Kotner et al. ⁴¹	1972	Case series
Kramer and Gatenby ⁴²	1996	Case report
Lindberg ⁴³	1966	Case series
Liu et al. ⁴⁴	2010	Case report
Lomeo et al. ⁴⁵	2007	Case report

(Continued)

TABLE 1. Continued

Author	Year	Study type
Losa et al. ⁴⁶	1992	Case report
McQuiston and Jones ⁴⁷	1959	Case report
Medini et al. ⁴⁸	1980	Retrospective review
Megat Shiraz et al. ⁷²	2008	Case report
Meziane et al. ⁴⁹	2012	Case report
Michalaki et al. ⁵⁰	2003	Retrospective review
Micozkadioglu et al. ⁵¹	2009	Case report
Miller et al. ¹¹	1998	Retrospective review
Natt and O'Sullivan ⁵²	2009	Case report
Noorani ⁵³	1975	Case series
Palacios et al. ⁵⁴	2002	Case report
Park et al. ⁵⁵	2009	Case report
Rainer ⁵⁶	1970	Case report
Schabel et al. ⁵⁷	1978	Case series
Schoeman et al. ⁵⁸	1997	Case report
Seltzer ⁵⁹	1974	Case report
Shahrizal et al. ⁷³	2009	Case report
Sharma et al. ⁶⁰	2002	Case report
Shih et al. ⁶¹	1995	Clinical investigation
Shreif et al. ⁶²	2001	Case report
Singh et al. ⁶³	1979	Case series
Thomas ⁶⁴	1965	Case report
Todd ⁶⁵	1965	Case report
Touma ⁶⁶	1971	Case report
Van Wart et al. ⁶⁷	1973	Case report
Vedrine et al. ⁶⁸	2009	Case series
Windfuhr and Ott ⁶⁹	2002	Case report
Yamaguchi et al. ²	2004	Case series
Zuo et al. ⁷⁰	2011	Retrospective review

that nasal obstruction was the most common presenting symptom; however, it was only noted in 29.8% of cases. Other commonly cited presenting symptoms included epistaxis (24.2%), facial swelling (9.9%), facial pain (9.9%), painless mass (6.8%), and change or loss of vision (6.2%). The highly variable symptomatology seen with our results is likely due to the anatomic intricacy where these tumors arise. Based on the location, a small increase in the size of a mass could create drastic differences in neural, vascular, or osseous damage.

The development of MM is a known sequelae of EMP, although controversy exists as to whether EMP and MM

TABLE 2. Summary of individual cases*

Characteristics	
Cases, n	175
Demographics	
Age, years, mean (range)	55 (5–79)
Gender	
Male	111 (69.4)
Female	49 (30.6)
Follow-up, months, mean; median (range)	60.9; 39 (2–288)
Presenting symptoms	
Obstruction	48 (29.8)
Epistaxis	39 (24.2)
Facial swelling	16 (9.9)
Facial pain	16 (9.9)
Painless mass	11 (6.8)
Change/loss of vision	10 (6.2)
Nasal discharge	7 (4.3)
CN VI palsy	6 (3.7)
Proptosis/ptosis	4 (2.5)
Headache	4 (2.5)
Anatomic site	
Nasal cavity and septum	63 (32.5)
Maxillary sinus and antrum	52 (26.8)
Nasopharynx	36 (18.6)
Ethmoid sinus	14 (7.2)
Sphenoid sinus	13 (6.7)
Paranasal sinus	12 (6.2)
Frontal sinus	4 (2.1)
Outcome	
ANED	100 (57.5)
AWD	25 (14.4)
DOD	24 (13.8)
D	24 (13.8)
Alive, unspecified	1 (0.6)
Follow-up measures	
Recurrence	28 (16)
Multiple myeloma	16 (9.1)

*Values are n (%) except where indicated.

ANED = alive, no evidence of disease; AWD = alive with disease; CN VI = 6th cranial nerve; D = died, other causes; DOD = died of disease.

TABLE 3. Frequency of using various treatment modalities in sinonasal plasmacytoma

Treatment	Cases (n)	% Total cases
Radiotherapy alone	89	50.9
Surgery alone	25	14.3
Chemotherapy alone	3	1.7
Surgery and radiotherapy	38	21.7
Radiotherapy and chemotherapy	9	5.1
Surgery and chemotherapy	0	0
Radiotherapy, surgery, and chemotherapy	6	3.4
No treatment	5	2.9

are different phases of the same disease or 2 different clinical entities.⁷⁵ The rate of conversion of EMP to MM is lower than other plasma cell neoplasms, such as SBP, with rates reported to be between 11% to 33% over 10 years.⁷⁶ The highest risk of conversion is reported to be in the first 2 years after diagnosis, but conversion has been noted up to 15 years after diagnosis.⁷⁵ Once conversion to MM is complete, less than 10% of patients will survive 10 years,^{74,75} although there is some evidence to suggest that patients with converted MM have a better prognosis than those that directly developed MM.^{11,75} In our review, 16 patients (9.1%) converted to MM, with a median follow-up of 39 months (3.25 years). Eleven of the 16 patients (68.8%) died of their disease. The rate of conversion to MM that we report is less than the expected 11% to 33% range, which could be attributed to the relatively short median follow-up in our review (3.25 years compared to the 10-year at-risk period for conversion). It is recommended that patients receive regular screening for MM every 6 weeks for the first 6 months after diagnosis of EMP and periodically thereafter due to the relatively high risk for conversion.¹⁰

Due to the rarity of the tumor, the lack of randomized clinical trials, and a propensity toward case reports, a debate continues to exist regarding the optimal management of solitary EMPs of the sinonasal tract. EMPs are radiosensitive tumors, so radiotherapy has traditionally been the mainstay of treatment.^{9,73} Local control rates of 80% to 100% have been reported with moderate-dose radiotherapy, with the optimal radiation dose between 40 and 50 Gy.^{9,71} Surgery is indicated for tumors that are localized and can be excised easily, although this is often difficult for EMPs originating in the sinonasal tract, due to the proximity to vital structures and the risk of poor cosmetic results.⁹ However, when clear surgical margins are obtained, the rate of local control with surgery alone is similar to that achieved with radiotherapy alone.⁷¹ Radiotherapy for reduction in tumor volume, followed by surgical excision, is often employed to reduce the invasiveness of surgery.⁹ A combination of radiotherapy and surgery can also be employed as salvage therapy for

TABLE 4. Treatment outcomes based on treatment modality used in sinonasal plasmacytoma

Outcome	Cases per treatment modality (n)	Cases per treatment modality (%)	Cases developing multiple myeloma (n)
Radiation alone	89		
ANED	55	61.8	0
AWD	7	7.9	2
DOD	13	14.6	10
D	13	14.6	0
Surgery alone	25		
ANED	14	56	0
AWD	3	12	0
DOD	3	12	0
D	4	16	0
Chemotherapy alone	3		
ANED	1	33.3	0
AWD	1	33.3	0
DOD	1	33.3	0
D	0	0	0
Surgery, radiation	38		
ANED	25	65.8	0
AWD	6	15.8	0
DOD	3	7.9	0
D	4	10.5	0
Radiation, chemotherapy	9		
ANED	4	44.4	0
AWD	4	44.4	1
DOD	1	11.1	0
D	0	0	0
Radiation, chemotherapy, surgery	6		
ANED	1	16.7	0
AWD	2	33.3	0
DOD	1	16.7	0
D	2	33.3	1
No treatment	5		
ANED	0	0	0
AWD	2	40	1
DOD	2	40	0
D	1	20	0

ANED = alive, no evidence of disease; AWD = alive with disease; D = died, other causes; DOD = died of disease.

recurrent disease.⁷³ Chemotherapy is not considered a first-line therapy for sinonasal EMPs as it improves neither local control nor overall survival, and can be associated with significant morbidity.⁷¹ Chemotherapy regimens are typically reserved for disseminated disease,⁷³ as a second-line treatment option for recurrent disease,⁹ or to delay the conversion to MM,^{1,71} although even these indications are controversial.

EMPs tend to have favorable outcomes when compared to other plasma cell neoplasms. Overall survival rate at 10 years is estimated to be 70%.⁷⁷ Alexiou et al.,⁷⁴ in a detailed, large-scale review of EMPs of the head and neck found that a combination of radiotherapy and surgery had the best 5-year survival rates by a significant margin. However, this study included cases from nearly an entire century (1905–1997), when diagnosis and treatment management may have changed drastically. Sasaki et al.,⁹ in their study of the efficacy of radiotherapy in the treatment of solitary EMPs of the head and neck, found that patients receiving any combination of radiotherapy and surgery had significantly better overall survival than patients who received radiotherapy alone or surgery alone. In their review of EMPs arising in the head and neck, Gerry and Lentsch⁷⁷ found that patients had a better response to surgical resection, whether used in combination with radiotherapy or as a single treatment modality.

Our review found that patients receiving a combination of radiotherapy and chemotherapy had the best outcomes, with 88.9% of these patients being classified as ANED or AWD. However, this subgroup consisted of only 9 patients. In our review, of the 3 most common treatment modalities (radiation alone, surgery alone, and combination surgery and radiotherapy), patients receiving a combination of radiotherapy and surgery had the best outcomes. This is consistent with the published literature, which indicates that overall survival is best for patients receiving combination therapy. Despite this finding, the most common treatment modality in this review was radiotherapy alone, for cases published both before and after 1990. In cases published before 1990, a combination of radiotherapy and surgery was the second most common treatment modality. For cases from 1991 and onward, surgery alone and combination radiotherapy and surgery were tied as the second most common treatment modalities.

Radiotherapy alone is likely the mainstay of treatment for many patients due to the marked radiosensitivity. However, as surgical technique, technology, and imaging improve, surgical management of sinonasal EMPs should be considered to improve overall outcomes.

The shift toward surgical management of small tumors should also be considered in light of our finding that the majority of patients that developed MM received radiotherapy alone as the initial management of the sinonasal EMP. This is consistent with Alexiou et al.'s⁷⁴ finding that patients receiving radiotherapy for solitary EMP of the head and neck had a higher conversion to MM. Although conversion rates to MM are lower for patients diagnosed

with EMP as compared to patients with SBP, all risk factors for conversion should be avoided. Due to the rarity of the tumor, its unique and complex location, and the potential morbidities associated with each treatment modality, management of sinonasal EMPs should be considered on a case-by-case basis. Factors such as tumor size, tumor location, risk for conversion to MM, degree of dissemination, and histopathological grade should be considered when determining the most appropriate treatment regimen.

Limitations

The limitations of this study are inherent to that of any systematic review. First, inherent to all pooled data analysis, there is weakness due to allocation and selection bias. Furthermore, there was a lack of uniformity in the reporting of information, especially including stage, grade, treatment modality, and outcomes, which limits our ability to perform a rigorous statistical analysis of the pooled data. We recognize that our inclusion of mainly case reports and case series increases the risk of bias. However, there is a lack of randomized controlled trials of sinonasal EMP, likely due to the rarity of the tumor, so these studies did not exist for

inclusion. We included cases from as early as 1955 in order to encompass as many cases as possible.

Conclusion

This review contains the largest pool of sinonasal EMP patients to date. Our findings suggest that radiotherapy alone is the most common type of treatment used, followed by combination surgery and radiotherapy, and surgery alone. Combination radiotherapy and chemotherapy treatment had the best outcomes in terms of survival and eradication of disease; however, this subgroup consisted of only 9 patients. Of the 3 most common treatment modalities, a combination of surgery and radiotherapy had the best survival outcomes. Conversion to MM occurred in 9.1% of patients, and those receiving radiotherapy alone had the highest conversion rate. Most of the data included in this study was extracted from case reports and case series. Further research on the optimal treatment modality, if feasible, should be done using randomized controlled clinical trials to ensure less bias and increased accuracy of results. 🌐

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