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SMARCB1-deficient sinonasal carcinoma: an updated systematic review and survival analysis

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Introduction/Objectives

- SWItch/Sucrose Non-Fermentable-related matrix associated actin-dependent regulator of chromatin subfamily B member 1 (SMARCB1)-Deficient Sinonasal Carcinoma (SDSC) is a rare malignancy of the nasal cavity and paranasal sinuses, accounting for approximately 3-5% of all sinonasal carcinomas.
- Our study aims to perform an updated systematic review and survival analysis on SDSC, to more accurately identify the clinicopathological features of this disease, as well as to determine survival outcomes and treatment effectiveness.

Materials and Methods

- References were identified through the search of five databases: PubMed, Cochrane, Embase, Web of Science and Scopus; from inception until 24th May 2025.
- Extracted information include cohort size, age, gender, clinical presentation, TNM staging, tumour site, microscopic features, immunohistochemical findings, genetic testing, follow-up duration, treatment modalities, survival outcome and status.
- We included two patients with SMARCB1-deficient sinonasal carcinoma treated at our institutions under the National Healthcare Group in the subsequent analysis.

Results

- A total of 646 studies were identified from the database, of which 70 studies were included in this review.
- 66 of the included studies with individual patient data were included in the survival analysis.

Baseline Characteristics and Clinical Presentation

- Median age was 54 years old (range 10-95), with 64.8% male and 34.7% female.
- The most common group of symptoms were nasal symptoms (n=97 25.5%), with the most common symptom being epistaxis (n=52, 14.0%), followed by nasal obstruction (n=36, 9.7%).

Treatment Outcomes

- Most patients were treated by surgery with concurrent or adjuvant chemotherapy and/or radiotherapy (n=148).
- Neoadjuvant chemotherapy in conjunction with surgery and adjuvant therapy was utilised in 27 patients.
- A total of 60 patients were treated non-surgically with either chemotherapy or radiotherapy or a combination of both.

Survival Analysis

- Median follow up duration was 14.5 months (range 0.3-180), of which 32.5% of patients died.
- Patients who received multimodality treatment had a statistically significant longer overall survival(OS) of 42 months (95% CI 27-81.4, p<0.001) compared to patients who only received unimodality treatment.
- Multivariable analysis of prognostic factors for SDSC showed that **N+ disease was a significant poor prognostic factor** for OS(HR=0.31, 95% CI 0.16-0.59, p<0.001), while **multimodality treatment** (HR=0.31, 95% CI 0.16-0.59, p<0.001) and **surgical treatment** (HR=0.50, 95% CI 0.26-0.96, p=0.04) were **favourable prognostic factors**.

Primary Tumour Site	No. of patients (percentage)
Nasal cavity/septum	139 (37.4%)
Ethmoid sinus	124 (33.3%)
Maxillary sinus	74 (19.9%)
Frontal sinus	28 (7.5%)
Paranasal sinuses (unspecified)	20 (5.4%)

Table 1: Breakdown of primary tumour sites for SDSC

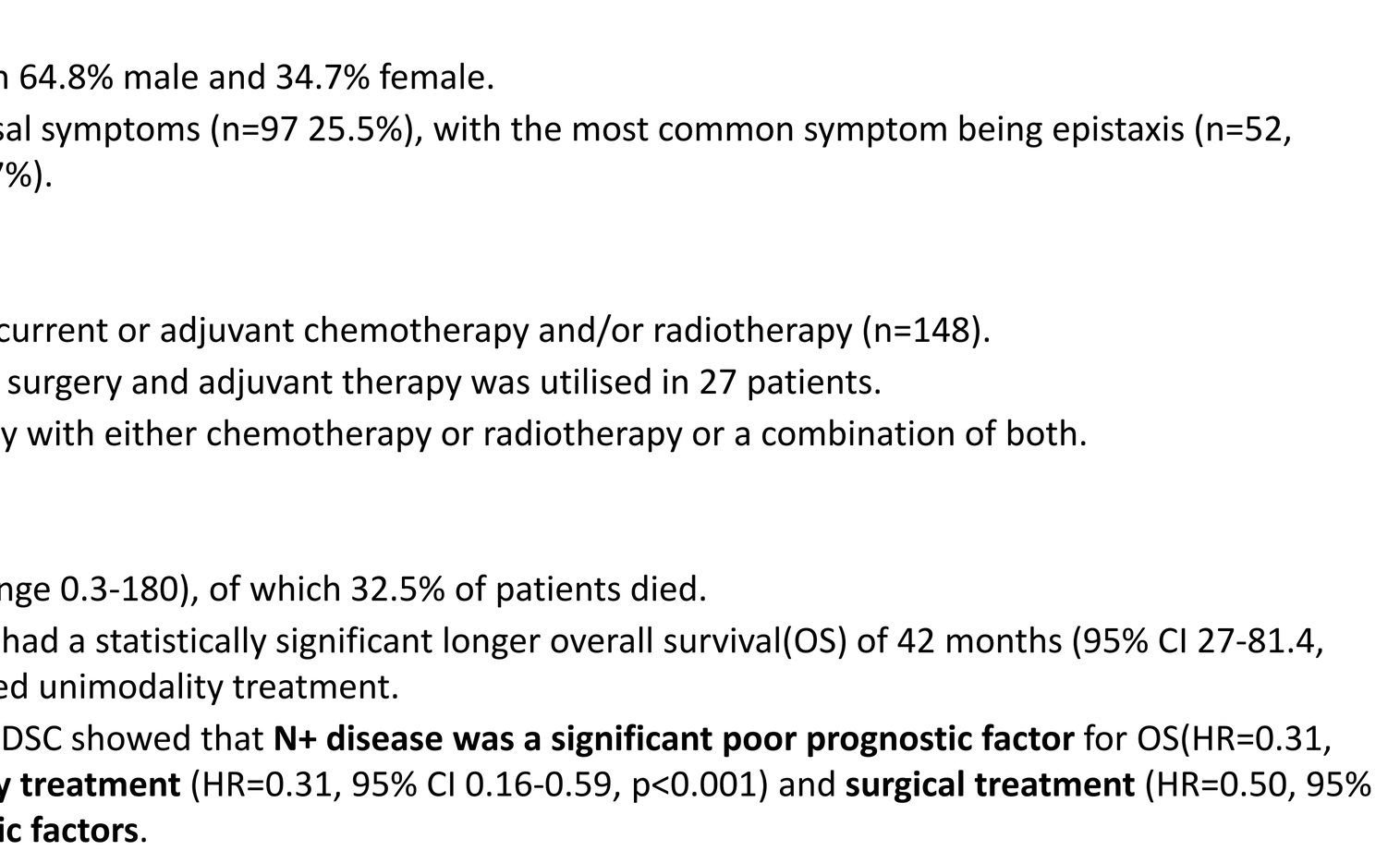

Figure 1: Kaplan-Meier curve showing 3-year overall survival

Conclusion

- N+ disease is a significant poor prognostic factor for SDSC.
- Patients who undergo surgical treatment and multimodality treatment have significantly better prognosis overall.