The Journal of Laryngology & Otology

cambridge.org/jlo

Sinonasal presentations of Rosai–Dorfman disease: a scoping review

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Review Article

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Cite this article: Ofori KA, Rimmer RA. Sinonasal presentations of Rosai–Dorfman disease: a scoping review. *J Laryngol Otol* 2025;1–6. https://doi.org/10.1017/S0022215125103046

Received: 14 June 2025
Revised: 7 July 2025
Accepted: 22 July 2025

Keywords:
histiocytosis; sinus; nasal cavity; paranasal sinuses; nasal obstruction; epistaxis; immunohistochemistry; tomography; X-ray computed; positron emission tomography computed tomography

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Abstract

Objectives. Rosai–Dorfman disease is a rare histiocytic disorder typically presenting with cervical lymphadenopathy. Sinonasal involvement is uncommon and presents diagnostic and therapeutic challenges. This scoping review synthesises literature on the clinical presentation, diagnosis, management and outcomes of sinonasal Rosai–Dorfman disease.

Method. We systematically searched PubMed, Scopus and Embase. Articles were screened using Endnote. Studies reporting sinonasal Rosai–Dorfman disease were included. The review followed Preferred Reporting Items for Systematic Reviews and Meta-Analyses-ScR (Scoping Review) guidelines.

Results. Thirty studies comprising 36 patients were included. Common symptoms were nasal obstruction (80.6 per cent) and epistaxis (41.7 per cent). Computed tomography (75 per cent) and magnetic resonance imaging (36.1 per cent) were primary imaging modalities. Histopathology showed emperipolesis (66.7 per cent), S-100 (69.4 per cent) and CD68 (47.2 per cent) positivity. Management was mainly surgical (72.2 per cent), with corticosteroids (44.4 per cent), radiotherapy (5.6 per cent) and chemotherapy (5.6 per cent) used less frequently. Outcomes included complete resolution (38.9 per cent), stable disease (38.9 per cent) and recurrence (16.7 per cent).

Conclusion. Diagnosis relies on histopathology and imaging. Surgical procedures, often with corticosteroids, remain the primary treatment. Future research should guide diagnostic and treatment protocols.

Introduction

Rosai–Dorfman disease is a rare histiocytic disorder first described in 1965 by Pierre Louis Destombes.[1] Histiocytoses are rare conditions involving the accumulation of cells such as macrophages and dendritic cells.[2] According to the 2016 revised histiocytosis classification, Rosai–Dorfman disease is under the 'R' group, encompassing its classical, sporadic and extranodal forms, and including variants associated with immune-mediated conditions such as systemic lupus erythematosus and neoplasia.[2] This classification reflects the clinical heterogeneity of Rosai–Dorfman disease and distinguishes it by its hallmark histiocytes with emperipolesis and S-100 positivity without CD1a expression.[2]

Rosai–Dorfman disease affects approximately 1 in 200 000 individuals, with about 100 new cases reported annually in the USA.[3] It frequently presents as bilateral cervical lymphadenopathy in children and adults.[4] Classical sporadic Rosai–Dorfman disease affects the lymph nodes and is more common in children and young adults, especially males of African descent.[4] Painless cervical lymphadenopathy, which is present in 80 per cent of cases, is often accompanied by systemic symptoms, including fever, night sweats, fatigue and weight loss.[2,5] Other lymph node sites may be involved.[2]

Rosai–Dorfman disease clinical behaviour encompasses moderate, disseminated and occasionally life-threatening forms.[2] Mild to moderate presentations often include isolated, painless cervical lymphadenopathy with or without systemic symptoms such as fever and night sweats. Disseminated disease may involve multiple extranodal sites, including the skin, orbit or paranasal sinuses.[2] Life-threatening cases may result from mass effect on the airway, brain or spinal cord.[6,7] Extranodal disease occurs in about 43 per cent of cases, affecting sites such as the skin, central nervous system and orbit.[1] Sinonasal involvement is rare, present in 11 per cent of cases and can be diagnostically challenging because of its similarity to other conditions.[8,9] Rosai–Dorfman disease-like histiocytes identified in chronic rhinosinusitis have prompted debate over classification.[9] Our scoping review aimed to synthesise current knowledge on the presentation, diagnosis and management of sinonasal Rosai–Dorfman disease, identifying gaps to inform future research and improve clinical practice.

Materials and methods

This scoping review adhered to the followed Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA)-ScR guidelines and involved a structured search of three databases, PubMed, Scopus and Embase, to gather relevant literature on sinonasal Rosai–Dorfman disease. These databases were chosen for their ability to index rare diseases and offer comprehensive coverage of biomedical and multidisciplinary literature.

The search targeted articles on sinonasal Rosai–Dorfman disease with the keywords 'Rosai–Dorfman disease', 'sinonasal', 'sinus', 'nasal cavity', 'paranasal sinuses' and 'histiocytosis' using Boolean operators (AND, OR) to refine the results. The complete search strategy can be found in Appendix 1.

Titles and abstracts were reviewed to identify potentially relevant studies and full-text reviews were conducted for articles meeting the inclusion criteria. Selected papers were then imported into EndNote for reference management and deduplication. Two reviewers independently screened and assessed the articles, and discrepancies were resolved through discussion to minimise bias.

Studies were included if the full text could be reviewed in English, reported a case or case series of sinonasal Rosai–Dorfman disease and included data on clinical features, diagnostic approaches or treatment strategies. Studies were excluded if they focused solely on nodal or non-sinonasal Rosai–Dorfman disease, were reviews, editorials or conference abstracts without detailed case data, or were non-English publications.

We created a table to extract key information from each study, including patient demographics, clinical presentation, diagnostic tools, treatment modalities and outcomes. Findings were synthesised descriptively to identify patterns in the presentation, diagnosis and treatment of sinonasal Rosai–Dorfman disease. Where appropriate, summary statistics and visual aids such as tables and figures were used to highlight trends and variations.

The Joanna Briggs Institute Critical Appraisal Tool was used to assess study quality in line with PRISMA-ScR, supporting the review's exploratory aims.

Results and analysis

A total of 154 articles were screened, with 30 studies (25 case reports and 5 case series) meeting inclusion criteria, comprising 36 patients with sinonasal Rosai–Dorfman disease.[5,8,10–37] Patients' ages ranged from 6 to 78 years (mean, 40.8**[Qwas.6]** ± 19.4 years), with a slight male preponderance (52.8 per cent) (Table 1). Most cases were reported in adults and seven paediatric cases were identified. Studies originated from 16 countries, with the USA (7 studies), India (6 studies) and Hong Kong (5 studies) contributing the most.

Nasal obstruction was the most common symptom, reported in 29 cases (80.6 per cent), followed by epistaxis (15 cases, 41.7 per cent), headache (7 cases, 19.4 per cent), sleep-disordered breathing symptoms (6 cases, 16.7 per cent), dyspnoea (5 cases, 13.9 per cent) and visual disturbances (4 cases, 11.1 per cent) (Table 2). Cervical lymphadenopathy was present in 14 cases (38.9 per cent).

All patients underwent diagnostic evaluation using multiple tools, with histopathological analysis typically based on sinonasal tissue and, in some cases, fine needle aspiration cytology (FNAC) of cervical lymph nodes. Immunohistochemical testing commonly included S-100, CD68 and immunoglobulin levels, while lymphocyte markers such as CD1a, CD3 and CD20 were used to exclude hematologic malignancies. Tissue sampling methods ranged from in-clinic biopsy to endoscopic sinus surgery and craniotomy. Emperipolesis is a non-destructive process in which a viable cell transiently resides within another living cell without structural or functional harm to either.[38] It is a hallmark feature of Rosai–Dorfman disease, typically accompanied by S-100 and CD68 positivity, key diagnostic features reported in 66.7, 69.4 and 47.2 per cent of cases, respectively.

Table 1. Patient characteristics

Characteristics	Value (*n* (%))
Total patients	36
Male	19 (52.8)
Female	17 (47.2)
Age (mean ± SD; years)	40.8 ± 19.4
Age range (years)	6–78

SD = standard deviation.

Table 2. Patient clinical presentations/symptoms

Symptom	Number of cases ($N = 36$)	%
Nasal obstruction	29	80.6
Epistaxis	15	41.7
Headache	7	19.4
Sleep-disordered breathing symptoms	6	16.7
Dyspnoea	5	13.9
Visual disturbances	4	11.1

All cases with available immunohistochemical data were positive for at least one of S-100 or CD68. Elevated immunoglobulin levels (IgG, IgA or IgG4) were observed in 6 cases (16.7 per cent). CD1a, CD3, CD20 and CD138 were each positive in one case, while all other lymphoma markers were negative. Erythrocyte sedimentation rate was documented in 11 cases (30.6 per cent), with only two normal results.

Computed tomography (CT) scans were used to evaluate disease in 27 cases (75 per cent) and magnetic resonance imaging (MRI) in 13 cases (36.1 per cent), with 5 patients receiving both. Positron emission tomography CT (PET-CT) was used in 5 cases (13.9 per cent) and 1 case utilised all three modalities. Imaging was performed in all but one case. Figure 1 illustrates the distribution of imaging modalities in our study.

Management involved multidisciplinary teams, including otolaryngology, pathology, radiology, medical oncology and radiation oncology. Treatment strategies varied by disease extent and severity. Surgical intervention was the most common approach, performed in 26 cases (72.2 per cent), using techniques such as endoscopic sinus surgery, carbon dioxide (CO_2) laser vaporisation and craniotomy. Corticosteroids were used in 16 cases (44.4 per cent), including 8 as adjuncts to surgery. Radiotherapy was used in two cases and chemotherapy was administered in two cases with systemic disease. Observation without active treatment was selected in 3 cases (8.3 per cent), some of which showed spontaneous regression. Figure 2 shows the distribution of treatment modalities.

Recurrence was reported in six cases, with varied timelines and management strategies. One patient developed pericardial involvement three years after sinonasal surgery, responding to

https://doi.org/10.1017/S0022215125103046

Figure 1. Venn diagram illustrating the use and overlap of imaging modalities in the diagnosis of sinonasal Rosai–Dorfman disease. CT = computed tomography; MRI = magnetic resonance imaging; PET-CT = positron emission tomography.

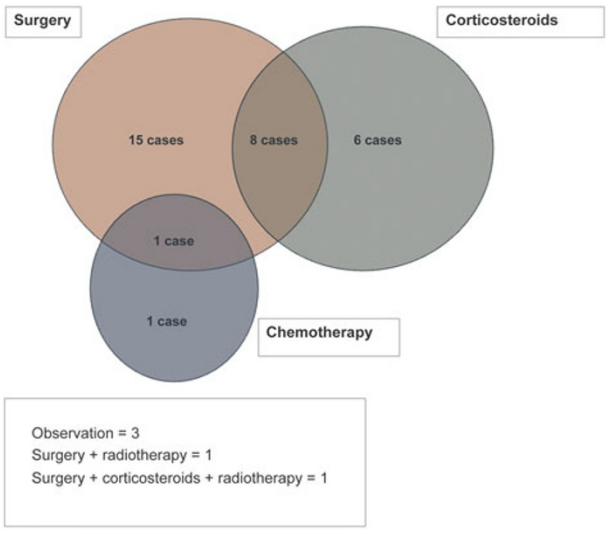

Figure 2. Venn diagram showing the overlap of treatment strategies used across sinonasal Rosai–Dorfman disease cases, including surgery, corticosteroids, chemotherapy and radiotherapy.

cladribine after methotrexate failure.[19] Another patient experienced recurrence within six months after surgical resection of extensive sinonasal and orbital disease and was subsequently lost to follow up.[24] In another case, a subglottic mass recurred following CO_2 laser debulking, with persistent sinonasal lesions and lymphadenopathy.[29] A patient with coexisting rhinoscleroma had surgical excision via the Caldwell–Luc approach but experienced minimal sinonasal recurrence, which was effectively managed with endoscopic sinus surgery.[29] A 72-year-old woman required repeat laser sessions within 3 months, achieving disease control by 10 months.[11] Finally, a 54-year-old man experienced symptom recurrence 3 years post-surgery but achieved sustained remission following systemic steroid therapy, with no recurrence during 3-year follow up.[36]

Among the reported cases, 14 patients (38.9 per cent) achieved complete resolution, while another 14 (38.9 per cent) had stable disease, including 1 with treatment-limiting complications due to immunosuppression. Recurrence occurred in 6 cases (16.7 per cent), often within 6 months post-treatment. Two patients (5.6 per cent) lacked follow-up data. Follow-up durations ranged from six weeks to five years.

Discussion

Rosai–Dorfman disease is considered a benign histiocytic disease typically presenting with massive, bilateral cervical lymphadenopathy, but sinonasal manifestations are uncommon.[8,23] Diagnosis at index presentation appears to be a challenge, often attributed to the presence of nonspecific sinonasal complaints, rarity and histological similarities to other diseases.[39,40]

Cervical lymphadenopathy was present in over a third of sinonasal Rosai–Dorfman disease cases and is frequently misdiagnosed as infection, granulomatous disease or malignancy.[39] Reports include a 45-year-old woman initially diagnosed with odontogenic infection, a 33-year-old man misdiagnosed with tuberculosis and fungal sinusitis, and a 45-year-old man misdiagnosed with lymphoma, all later confirmed to have Rosai–Dorfman disease.[41–43] Rosai–Dorfman disease can be differentiated from Langerhans cell histiocytosis by emperipolesis and its characteristic immunophenotype, $CD68^{+}$/$S\text{-}100^{+}$/$CD1a^{-}$.[2,44,45] Rarely, the disease may extend intracranially and mimic meningioma, but its hallmark histologic features aid differentiation, underscoring the need for heightened diagnostic awareness.[35,46,47]

Although FNAC may suggest Rosai–Dorfman disease, it is often inconclusive and can be confounded by benign mimics, thus definitive diagnosis typically requires tissue biopsy with detailed histopathological and immunohistochemical evaluation.[29,39,48,49]

Of the two cases tested for Epstein–Barr virus (EBV), one was positive. Rosai–Dorfman disease is thought to involve immune dysregulation triggered by an infectious agent, specifically the EBV.[39] Other implicated disease-causing pathogens include human herpesvirus (HHV)-6, Varicella zoster virus, Cytomegalovirus, *Klebsiella* spp. and *Brucella* spp.[11,50] However, the aetiology of Rosai–Dorfman disease remains unclear, and the role of EBV is not well-established, hence future research should systematically evaluate EBV status in Rosai–Dorfman disease patients to determine whether EBV-driven immune responses contribute to disease pathogenesis.[39,50]

Rosai–Dorfman disease is often multifocal, and diagnosis in one area should prompt suspicion of multi-system involvement.[51] Although ultrasonography is recommended as the initial imaging modality for children with neck masses, it may be insufficient in suspected Rosai–Dorfman disease cases.[52] A two-year-old girl with cervical lymphadenopathy had normal ultrasound findings, but CT imaging revealed a sinonasal mass with sphenoid sinus erosion and orbital extension, highlighting the critical role of comprehensive imaging in patient evaluation.[39,53]

Given Rosai–Dorfman disease's potential for systemic involvement, PET-CT is increasingly utilised to detect subclinical disease, particularly in patients with otherwise localised symptoms. Studies suggest PET-CT provides additional diagnostic information in 30 per cent of cases where CT or MRI is inconclusive, leading to management modifications in up to 41 per cent of patients.[34,54] In one sinonasal Rosai–Dorfman disease case, PET-CT detected skeletal involvement missed by prior imaging, prompting a change in treatment.[34] In another, a 42-year old woman with longstanding extranodal Rosai–Dorfman disease underwent PET-CT monitoring over nearly a decade, which revealed recurrent Fluorodeoxyglucose (FDG)-avid lesions and guided treatment with corticosteroids, radiotherapy and 2-chlorodeoxyadenosine, resulting in partial responses.[55] Routine

https://doi.org/10.1017/S0022215125103046

PET-CT should be considered in cases with suspected systemic involvement or unclear disease extent on conventional imaging.[54]

Treatment for Rosai–Dorfman disease is primarily indicated for symptomatic cases or those involving critical organ dysfunction.[40,56] While some cases resolve spontaneously, up to 70 per cent may persist.[35,57] Localised symptomatic disease is generally treated with surgery or radiotherapy, while asymptomatic cases are monitored.[56,57]

In our review, surgery was the most common treatment, particularly for resectable sinonasal disease. A systematic review of paediatric otorhinolaryngologic Rosai–Dorfman disease found surgical excision to be the most common and effective treatment, with the lowest recurrence rates.[47] However, several studies emphasise the value of combining medical and surgical therapies, and the importance of individualised treatment strategies.[57] If complete resection is not feasible, adjuvant chemotherapy or radiotherapy may support disease control.[13] Laser excision is a minimally invasive option for nasal lesions, while corticosteroids and endoscopic surgery are useful in recurrent or compressive cases. However, the optimal surgical approach remains undefined.[11]

Corticosteroids were suggested as the first-line treatment in several cases, but with limited efficacy in severe disease.[57] Studies suggest treatment with corticosteroids results in transient decrease in lymph node size but rebound frequently occurs after treatment.[13] In other studies, however, corticosteroids were shown to effectively reduce fever and lymphadenopathy in Rosai–Dorfman disease, with symptom resolution reported within a range of 5 days to 6 months.[13] These highlight the variability in responses, with some patients requiring prolonged therapy or experiencing relapse.[58] While corticosteroids are valuable for symptom control, long-term management may necessitate additional therapeutic approaches.

A patient in our review received methotrexate, vinblastine and 6-mercaptopurine (MP), indicating a potential role for immunosuppressive agents in select cases.[10] Additionally, a 64-year-old man with refractory, disseminated Rosai–Dorfman disease reported complete remission following treatment with siltuximab, an anti-interleukin (IL)-6 monoclonal antibody, with minimal adverse effects.[56] The role of IL-6 in Rosai–Dorfman disease pathophysiology suggests that cytokine dysregulation may contribute to disease progression.[56] While the lack of IL-6 measurement is a limitation, the favourable clinical response supports the need for further investigation. A separate review of seven Rosai–Dorfman disease patients, five with prior treatment failures, found that thalidomide led to clinical improvement in five cases and minimal response in two.[59] Thalidomide, an oral immunomodulatory agent, exerts anti-inflammatory effects through cytokine modulation, tumour necrosis factor-α inhibition and T-helper cell regulation.[59,60] However, its use is limited by significant adverse effects, including hypothyroidism, deep vein thrombosis and teratogenicity.[60]

Cases of widespread systemic Rosai–Dorfman disease often require multidisciplinary management involving surgical specialists, oncology, rheumatology and immunology. Radiotherapy may be considered for refractory or isolated lesions, but its overall effectiveness has been limited in prior reports.[61]

Rosai–Dorfman disease has an unpredictable disease course, with approximately 50 per cent of cases resolving spontaneously, 33 per cent showing residual asymptomatic lymphadenopathy and 17 per cent exhibiting persistent disease.[24] Clinical assessment and imaging should be performed every 6 to 12 months, particularly in patients with prior recurrence or incomplete resection. Surgical cases may require earlier imaging, often within three to six months, to assess for residual disease.

Long-term disease monitoring is essential, as highlighted by a case of sinonasal Rosai–Dorfman disease diagnosed in the 1990s that recurred over a decade later with symptomatic tracheobronchial obstruction requiring mechanical resection.[62] A clinical study of 10 sinonasal Rosai–Dorfman disease cases over 7 years reported a high recurrence rate, reinforcing the importance of extended surveillance.[63]

Asymptomatic or mild cases of Rosai–Dorfman disease may be managed with observation and routine follow-up, given the possibility of spontaneous resolution. For symptomatic, localised sinonasal disease, endoscopic surgery is often the preferred initial treatment. Corticosteroids may be useful in recurrence or diffuse inflammation, while refractory disease may require adjunctive therapies such as radiotherapy, chemotherapy or targeted agents like siltuximab, although evidence remains limited. In multifocal or systemic disease, PET-CT should guide treatment decisions, and a multidisciplinary approach involving surgery, oncology and immunology is advised. CT remains the best imaging modality for evaluating bony anatomy and disease extent.[64]

For post-surgical surveillance, endoscopy and imaging (MRI, CT or PET-CT) are advised, with MRI showing the highest positive predictive value.[65] According to National Comprehensive Cancer Network (NCCN) guidelines for sinonasal tumours, 80–90 per cent of recurrences occur within the first 2–4 years, so surveillance typically includes 4–12 visits in the first year, 3–6 in the second, 2–3 annually from years 3 to 5 and then either yearly follow up or discontinuation.[66] Studies suggest that up to one-third of follow-up visits may be unnecessary, highlighting the need for individualised surveillance.[66]

This review consolidates current knowledge on sinonasal Rosai–Dorfman disease, emphasising the diagnostic and therapeutic challenges associated with this rare condition. Future research should aim to refine diagnostic criteria, establish standardised treatment protocols and identify markers predictive of disease progression and recurrence.

Our findings are limited by the rarity of sinonasal Rosai–Dorfman disease, with most data drawn from retrospective case reports and small series. The absence of uniform diagnostic and treatment guidelines, along with inconsistent reporting of key clinical details, such as immunohistochemical findings, EBV status, follow-up duration and recurrence, hinders the ability to draw definitive conclusions. Only two cases included EBV testing, leaving its role in disease pathogenesis unclear. Additionally, long-term outcomes were often inadequately documented, making the true recurrence risk difficult to assess.

Larger, prospective studies with standardised reporting and extended follow up are essential to improve the clinical decision-making and long-term management of sinonasal Rosai–Dorfman disease.

- Sinonasal Rosai–Dorfman disease is rare and presents with non-specific symptoms that complicate timely diagnosis
- Surgical intervention, occasionally combined with corticosteroids, remains the primary treatment modality for symptomatic disease
- Histopathology showing emperipolesis, S-100$^+$/CD68$^+$ staining and imaging are essential for diagnosis
- PET-CT is increasingly valuable for assessing systemic involvement, tracking recurrence and treatment response
- Future research should focus on standardised diagnostic criteria, optimised therapies and long-term surveillance protocols

https://doi.org/10.1017/S0022215125103046

Conclusion

Sinonasal Rosai–Dorfman disease is a rare disease that remains challenging to diagnose and manage because of its variable presentation and lack of standardised guidelines. Diagnosis often requires a combination of tissue analysis and immunohistochemistry. Treatment varies widely, with surgery being the most common approach, often combined with corticosteroids, radiation or chemotherapy. PET-CT has proven useful in tracking disease progression and detecting recurrence, suggesting its potential role in long-term follow up. With the risk of recurrence and the absence of clear guidelines, future research should focus on refining diagnostic criteria, optimising treatment plans and developing structured follow-up protocols. Collaboration among specialists is essential to improving patient care and expanding our understanding of this uncommon but important disease.

Competing interests. None declared.

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Appendix 1. Search strategies

Pubmed

('Rosai-Dorfman disease'[MeSH Terms] OR 'Rosai-Dorfman disease'[Title/Abstract] OR 'Sinus histiocytosis with massive lymphadenopathy'[Title/Abstract] OR 'SHML'[Title/Abstract]) AND ('Sinonasal'[Title/Abstract] OR 'Head and Neck'[MeSH Terms] OR 'Nasal cavity'[MeSH Terms] OR 'Paranasal sinuses'[MeSH Terms] OR 'Ethmoid sinus'[Title/Abstract] OR 'Maxillary sinus'[Title/Abstract] OR 'Frontal sinus'[Title/Abstract] OR 'Sphenoid sinus'[Title/Abstract] OR 'Nasal mass'[Title/Abstract] OR 'Nasal discharge'[Title/Abstract] OR 'Nasal obstruction'[Title/Abstract] OR 'Rhinosinusitis'[Title/Abstract] OR 'Sinus tumor'[Title/Abstract])

Embase

('rosai-dorfman disease' OR 'sinus histiocytosis with massive lymphadenopathy') AND ('sinonasal' OR 'nasal cavity' OR 'paranasal sinus' OR 'frontal sinus' OR 'maxillary sinus' OR 'sphenoid sinus' OR 'ethmoid sinus' OR 'nasal obstruction' OR 'rhinosinusitis') NOT ('skin' OR 'orbital' OR 'bone' OR 'CNS' OR 'lymph nodes')

Scopus (Advanced search)

TITLE-ABS('Rosai-Dorfman disease' OR 'Sinus histiocytosis with massive lymphadenopathy' OR 'SHML') AND TITLE-ABS('sinonasal' OR 'nasal cavity' OR 'paranasal sinuses' OR 'ethmoid sinus' OR 'maxillary sinus' OR 'frontal sinus' OR 'sphenoid sinus' OR 'nasal mass' OR 'nasal obstruction' OR 'rhinosinusitis')

*Date of most recent search: 8 February 2025.

https://doi.org/10.1017/S0022215125103046