

Multimodal treatment of a low-grade Nasosinusal Sarcoma: Insights from a rare case

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Abstract

Biphenotypic sinonasal sarcoma (BSNS) is a rare, recently recognised malignant tumour of the sinonasal tract, characterised by distinctive biphenotypic myogenic and neural differentiation and frequently associated with the PAX3-MAML3 gene rearrangement. Its locally aggressive behaviour and proximity to critical structures pose significant diagnostic and therapeutic challenges. We report a case of a 59-year-old female presenting with a progressively enlarging intercanthal mass, ultimately diagnosed as BSNS with ethmoidal, orbital, and endocranial extension. Surgical resection was performed, including ethmoidectomy and removal of the frontal tumour component, achieving near-complete excision while preserving vital structures. Post-operative imaging revealed a small residual tumour in the left ethmoidal cells with extra-conical fat infiltration, necessitating adjuvant radiotherapy with a total dose of 70 Gy in 35 fractions. The treatment was well tolerated, with only mild acute side effects. At two-year follow-up, there was no evidence of local or loco-regional recurrence, highlighting the effectiveness of a combined modality approach in managing advanced BSNS. This case underscores the importance of a multidisciplinary strategy involving surgery and radiotherapy when complete resection is not feasible due to anatomical constraints. Clear patient counselling regarding recurrence risk and the necessity of regular long-term follow-up is essential to optimise outcomes. Our experience contributes to the limited literature on BSNS management, reinforcing current recommendations and advocating for further studies to establish standardised treatment protocols for this rare entity.

Keywords: Biphenotypic sinonasal sarcoma; Sinonasal sarcoma; Skull base neoplasm; Orbital invasion; Adjuvant radiotherapy; Multidisciplinary approach

1. Introduction

Biphenotypic sinonasal sarcoma (BSNS) is a recently recognised type of sarcoma arising exclusively in the sinonasal tract with a unique clinical course, histopathology and genetics. Due to its rarity, only case series and case reports are available¹. It most commonly presents with a PAX3-MAML3 rearrangement^{2,3}. Its treatment usually involves a combination of surgery, radiotherapy and chemotherapy. Understanding this disease requires a multidisciplinary approach involving ENT specialists, oncologists and radiologists.

We report here a case of locally advanced biphenotypic nasosinusal sarcoma with endocranial extension, managed with combined therapy, and whose two-year follow-up showed no sign of recurrence.

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2. Case report

We report the case of a 59-year-old female patient, with no notable medical or surgical history, who consulted for a chronically evolving intercanthal mass, progressively increasing in size, with no associated functional signs.

2.1. Clinical examination

The ENT examination revealed a firm, painless intercanthal mass, fixed in relation to the deep plane, with normal-looking skin opposite. Decreased left nasal flow was noted with no other obstructive signs.

2.2. Surgical management

Removal of the endonasal portion of the tumour was performed, combined with a left ethmoidectomy and a middle meatotomy with drainage of the maxillary sinus. The frontal contingent of the mass was also removed. The dura was exposed, while respecting the adjacent anatomical structures, in particular the periorbit and the anterior ethmoidal artery.

2.3. Post-operative course and adjuvant treatment

Post-operative imaging revealed a tumour residue measuring 12 x 13 mm in the left ethmoidal cells, with invasion of the papyraceous lamina and infiltration of extraconical fat (Figure 2). Given the impossibility of revision surgery, adjuvant radiotherapy was started at a dose of 70 Gy in 35 fractions, after obtaining the patient's informed consent.

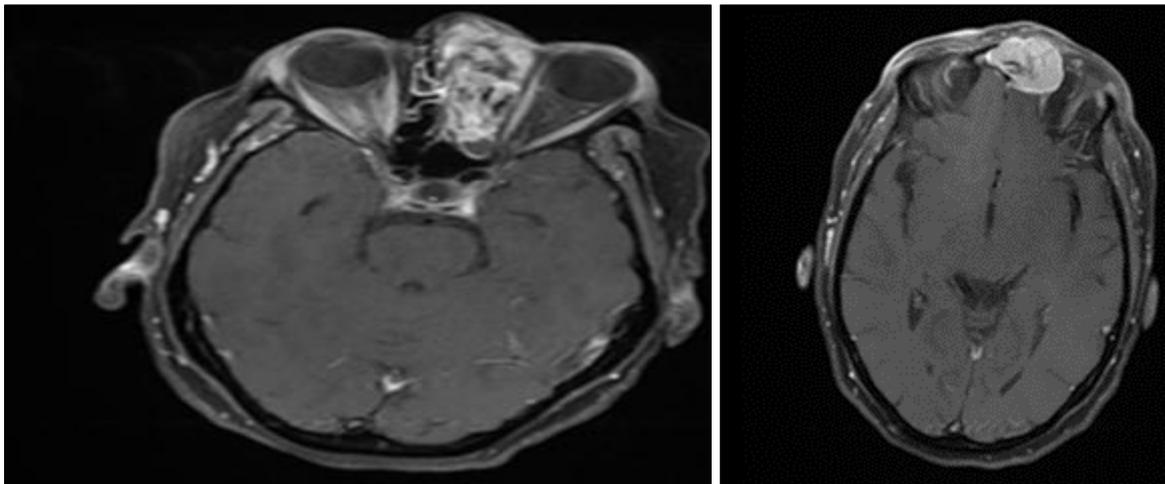


Figure 1 T1 gado craniofacial MRI: transverse section showing -Presence of a tissue process centred on the left ethmoidal cells, described as T1 hypointense, intensely enhancing after contrast, associated with thickening of the ethmoidal cell walls, with lysis of the roof of the orbit and endocranial extension, but limited by the dura mater

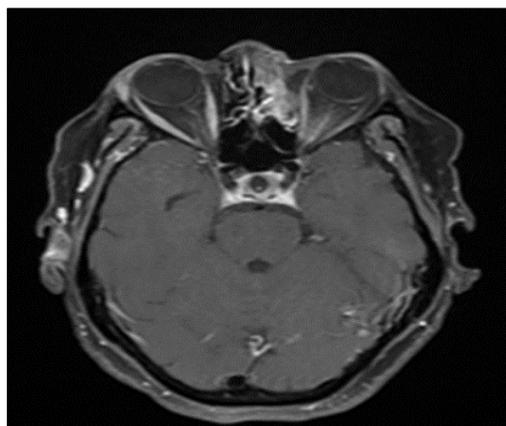


Figure 2 T1 gado sequence craniofacial MRI showing a small 12*13mm measured tumour residue at the level of the left ethmoidal cells responsible for an effraction of the papyraceous lamina with infiltration of the extra conical fat

Side effects were limited to grade I radiodermatitis, associated with lacrimation and conjunctival hyperhaemia, with no other notable acute complications.

2.3.1. Follow-up

Clinical and radiological assessment at 2 years post-treatment revealed no evidence of local or loco-regional recurrence or late complications.

3. Discussion

The case report describes a 59-year-old female patient with no prior history of locally advanced biphenotypic nasosinus sarcoma (BSNS) with endo-orbital and endocranial extension, a situation that illustrates the diagnostic and therapeutic complexity of this rare tumour. BSNS has recently been recognised as a distinct entity by the WHO [2] and is characterised by myogenic and neuronal differentiation, often associated with a PAX3-MAML3 translocation specific to this disease [3]. Clinically, this tumour manifests as an often painless, slow-growing nasal mass, but with locally aggressive infiltrative behaviour, which was the case in our patient [3,4]. Involvement of the ethmoidal cells, the orbit and erosion of the ethmoid roof with meningeal contact are classic extensions, although endocranial invasion beyond the dura remains rare [4,5].

The gold standard of treatment is wide surgical resection, with the aim of obtaining healthy margins [5]. However, in cases such as this, where the proximity of critical structures (meninges, orbit) limits the surgical procedure, a tumour residue may persist. Post-operative imaging confirmed a residue measuring 12×13 mm with infiltration of extra-conical fat, justifying adjuvant radiotherapy. This approach is supported by several studies which recommend radiotherapy in cases of positive margins, bone or dural invasion, or in cases where the tumour is not completely resectable [6-8]. Intensity modulated conformal radiotherapy (IMRT) is the preferred technique, providing optimal coverage while preserving sensitive structures [7]. In our case, a dose of 70 Gy in 35 fractions was administered, in line with current recommendations for positive margins or macroscopic residual [9].

In terms of tolerance, the patient experienced moderate acute side effects (grade I radiodermatitis, ocular redness, lacrimation), which are common in irradiation of the naso-orbital region [10]. These acute toxicities are generally well controlled with symptomatic care, although ophthalmological follow-up is recommended because of the risk of late ocular sequelae (keratitis, dryness, optic neuropathy) [11]. The absence of local recurrence or progression at 18 months underlines the efficacy of the combined surgery-radiotherapy strategy in locally advanced forms of BSNS. It should be noted that chemotherapy was not proposed here, in line with the literature which limits its indication to non-operable or metastatic cases, in the absence of a standardised protocol [12].

This case illustrates the challenges posed by BSNS extending to the base of the skull, requiring a multidisciplinary approach. It confirms the value of post-operative radiotherapy in incompletely resected forms, with a good locoregional response in the medium term, despite the potential morbidity of the treatment.

4. Conclusion

Nasosinus sarcoma is a rare and very aggressive tumour, with an often-unfavourable prognosis due to its tendency to recur. Treatment combines surgery, radiotherapy and sometimes chemotherapy. Clear communication with patients about the risk of recurrence and the importance of regular follow-up is essential. A multidisciplinary approach is crucial to optimise management and results.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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