

## CASE STUDY

# Synovial Sarcoma of the Nasal Cavity. A Case Report<sup>☆</sup>

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### KEYWORDS

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### PALABRAS CLAVE

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Sarcoma sinovial de  
fosas nasales;  
Sarcoma sinovial de  
cabeza y cuello

**Abstract** Synovial sarcoma is a rare tumour found in soft tissue; it is a mesenchymal spindle cell tumour that is not related to the synovial membrane. This tumour has a low incidence, the most frequent place of occurrence being the lower extremities in young adults. Synovial sarcoma of the head and neck accounts for 3%–5% of sarcomas in this anatomical region. The treatment of choice for synovial sarcoma of the head and neck is a complete surgical excision of the tumour mass followed by adjuvant radiotherapy.

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### Sarcoma sinovial de fosas nasales. A propósito de un caso

**Resumen** El sarcoma sinovial es una rara neoplasia encontrada en tejidos blandos; es un tumour mesenquimal de células fusiformes que no está relacionado con la membrana sinovial y muestra diferenciación epitelial variable. Típicamente surge en las extremidades, pero aunque se ha descrito en casi todas las localizaciones de cabeza y cuello, la afectación de esta región es mucho menos común.

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## Clinical Case

We present the case of a 49-year-old male who consulted for repeated epistaxis from both nostrils, bilateral nasal obstruction and blurred vision in his left eye of over 3 months' duration, without diplopia.

A nasal fibroscopy showed both nostrils were completely occupied by a mass of polypoid appearance, friable and prone to haemorrhage. The ophthalmologic examination

found no alterations of ocular motility or reflexes. Visual acuity only allowed fingers to be counted at 2 m (less than 0.1).

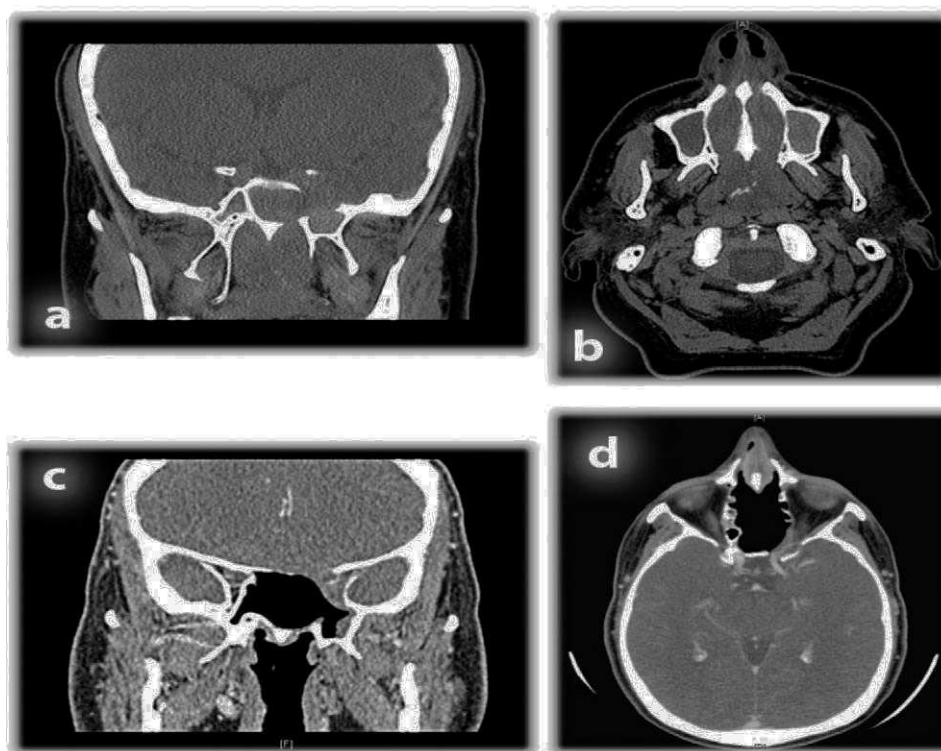
The cervicofacial CT (Fig. 1) revealed a mass occupying both nasal cavities, with destruction of the lamina papyracea of the left nostril and involvement of the left orbit, extending into the left cavernous sinus and both sphenoid sinuses. An MRI was not performed because the patient carried a fixed metal prosthesis incompatible with the test.

We performed a biopsy of the lesion by endoscopic sinonasal surgery. The microscopic study indicated a proliferation of predominantly spindle-type cells, with elongated, irregular and hyperchromatic nuclei, although they acquired a more epithelioid morphology in some pockets. There were areas of necrosis with figures of typical and atypical mitoses. In the immunohistochemical analysis the tumour

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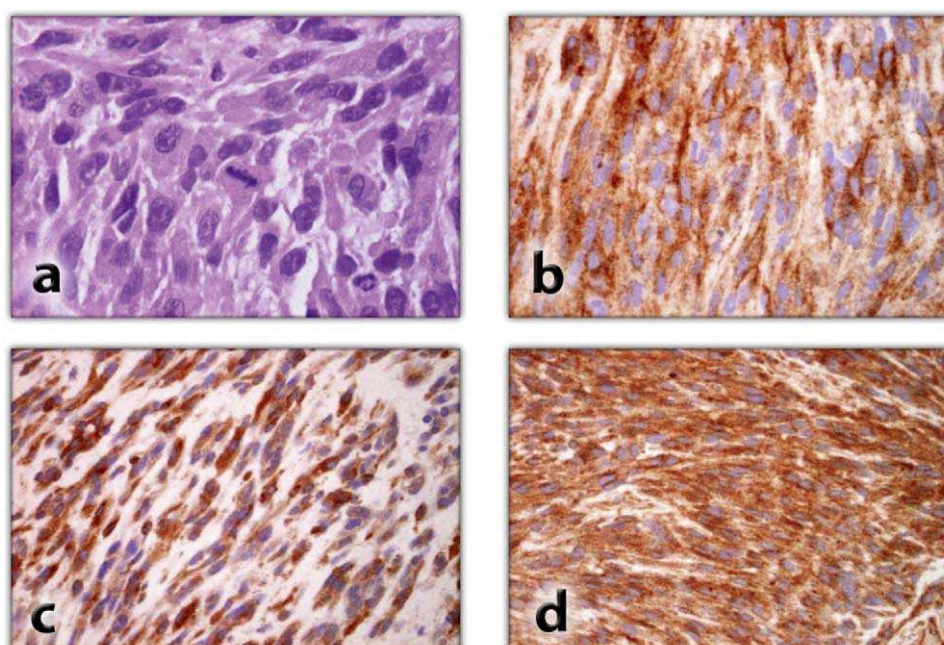
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**Figure 1** (a) and (b) Preoperative CAT images. (c) and (d) Postoperative CAT images, one year after treatment.

cells expressed immunoreactivity to epithelial membrane antigen, vimentin, PS-100, BCL-2, CD99, and CD56 (Fig. 2). The definitive diagnosis was poorly differentiated synovial sarcoma of the nasal fossa (two-phase version).

We performed a complete removal of the tumour mass, encompassing both ethmoids and the medial walls of both maxillary sinuses (not infiltrated, but occupied by a tumour mass that was also removed) and the rhinopharyngeal



**Figure 2** (a) Cellular mitosis (haematoxylin and eosin staining). (b) EMA (epithelial membrane antigen) immunoreactivity using immunohistochemical technique. (c) BCL-2 immunoreactivity using immunohistochemical technique. (d) CD99 immunoreactivity using immunohistochemical technique.

extension, along with the anterior and medial walls of both sphenoid sinuses. After removal, we noted a wide exposure of the anterior fossa dura mater. Both frontal recesses were free of tumour. Excision was completed by removal of the orbital extension of the tumour that affected part of the orbital fat without reaching the extrinsic musculature and the tumour remains in relation to the optic nerve, which was seemingly not infiltrated, but was devoid of its bony cover. The posterior portion of the nasal septum was removed from the sphenoid rostrum up to the anterior edge of the defect in the roof of the orbit.

Postoperative evolution included cures without incidences. The patient referred a subjective improvement of visual acuity in the hours following surgery.

Given the characteristics of the tumour, the patient underwent radiotherapy treatment, being administered a total dose of 50 Gy with fractionation of 200 Gy/d with 3 ELA photon fields of 6 MV.

Monthly checks were performed and the patient once again referred loss of visual acuity in his left eye after radiotherapy. We found atrophy of the optic disc and visual acuity that could sense light, project it and count fingers at 20 cm.

After 15 months of follow-up, there are no signs of recurrence or metastasis (Fig. 1).

## Discussion and Conclusions

In a study of 40 patients with synovial sarcoma of head and neck, Harb et al.<sup>1</sup> found that the most frequent location was the neck (60%) and that it was found even in relatively young patients (mean age of 29 years). The latter fact is supported in the literature consulted.<sup>2-9</sup>

Regarding histopathology, most authors report a preference of single-phase forms,<sup>1,3,4,10</sup> although this proportion is not significantly higher (58%) in long series.<sup>1</sup>

There have been reports of synovial sarcoma in almost all locations of the head and neck area, including the nasopharynx,<sup>10</sup> tonsils,<sup>6</sup> oropharynx,<sup>7</sup> thyroid gland<sup>8</sup> and parapharyngeal space.<sup>2</sup> Nasal and maxillary involvement<sup>11</sup> are particularly rare, with only some isolated cases being found.<sup>1,4,10</sup> The most frequent location is cervical,<sup>1,3,4</sup> including the parotid gland.<sup>4</sup> In the literature, there is an apparent relationship with a history of head and neck radiation.

Regarding treatment, the most common option was surgery followed by radiotherapy.<sup>1-3</sup> Local recurrences were described especially in relation to protocols without radiotherapy.<sup>3</sup>

The prognosis seems to depend on location (better in the upper aerodigestive tract),<sup>1</sup> size (better for those less than 5 cm)<sup>1</sup> and extension (better for those without bone extension).<sup>1</sup> There are various cases in the literature with a good outcome, which are free from disease after monitoring for many years.<sup>1,3</sup>

The present case is of particular rarity, as it occurred in a patient older than the average found in the literature, with a two-phase variant and a nasal location. After following the treatment proposed in the literature, the patient is currently free from tumour, 15 months after surgery.

Despite the low incidence of this neoplasm in the head and neck region (less than 5% of sarcomas in this area<sup>3</sup>) it should be included in the differential diagnosis in cases of masses in the intranasal region. However, the rarity of this location encourages the reporting of any cases found, in order to determine the clinical, epidemiological and prognostic characteristics of the tumour more accurately, as well as its appropriate treatment.

## Conflict of Interest

The authors have no conflicts of interest to declare.

## References

1. Harb WJ, Luna MA, Patel SR, Ballo MT, Roberts DB, Sturgis EM. Survival in patients with synovial sarcoma of the head and neck: association with tumor location, size, and extension. *Head Neck*. 2007;29:731-40.
2. Khademi B, Daneshbod Y, Negahban S, Daneshbod K, Kaviani M, Mohammadianpanah M, et al. Biphasic parapharyngeal synovial sarcoma: a cytologic and immunocytologic report of a case. *Cytojournal*. 2006;3:20.
3. Sefiani S, Amarti A, Boulaadas M, Maher M, Saidi A. Synovial sarcoma of the head and neck: two cases report. *Rev Laryngol Otol Rhinol (Bord)*. 2005;126:53-6.
4. Al-Daraji, Lasota J, Foss R, Mienttinent M. Synovial sarcoma involving the head: analysis of 36 cases with predilection to the parotid and temporal regions. *Am J Surg Pathol*. 2009. Epub ahead of print.
5. Sharif MA, Mushtaq S, Mamoon N, Khadim MT, Asghar Z. Biphasic synovial sarcoma of oral cavity. *J Coll Physicians Surg Pak*. 2008;18:731-5.
6. Ishiki H, Miyajima C, Nakao K, Asakage T, Sugawara M, Motoi T. Synovial sarcoma of the head and neck: rare case of the cervical metastasis. *Head Neck*. 2009;31:131-5.
7. Kuehnemund M, Friedrichs N, Bootz F. Synovial sarcoma of the head and neck. *Laryngorhinootologie*. 2008;87:498-502. Epub.
8. Jang KS, Min KW, Jang SH, Paik SS, Tae K, Jang SJ, et al. Primary synovial sarcoma of the thyroid gland. *J Korean Med Sci*. 2007;22 Suppl.:S154-8.
9. Orlandi E, Zonca G, Pignoli E, Stucchi C, Borroni M, Collini P, et al. Postoperative radiotherapy for synovial sarcoma of the head and neck during pregnancy: clinical and technical management and fetal dose estimates. *Tumori*. 2007;93:45-52.
10. Lin HC, Friedman M, Kuo FY, Huang HY. Synovial sarcoma of the Eustachian tube. *Head Neck*. 2009;31:1245-8.
11. Luo CW, Liu CJ, Chang KM. Synovial sarcoma of the temporomandibular joint area: report of a case. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2007;104:e62-5. Epub 2007 Aug 20.