

## Synovial sarcoma of the ethmoidal sinus

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### Abstract

**Background:** Synovial sarcoma is a high-grade, soft tissue, malignant disease associated with poor outcome. Typically, synovial sarcoma involves the extremities, with less than 10 per cent of cases occurring in the head and neck region. Synovial sarcoma of the paranasal sinuses is a rare entity. This paper presents a case of an elderly patient with synovial sarcoma of the ethmoidal sinus.

**Case report:** An 80-year-old woman who had right epistaxis underwent nasal endoscopy and biopsy. The pathology indicated synovial sarcoma and the patient underwent endoscopic excision of the tumour.

**Conclusion:** Synovial sarcoma of the ethmoidal sinus is very rare. Patients should undergo excision of the tumour with post-operative radiotherapy. However, the prognosis remains poor and usually the patient succumbs to death within a year.

**Key words:** Sarcoma, Synovial; Paranasal Sinuses

### Introduction

Synovial sarcoma is a high-grade, soft tissue, malignant disease associated with poor outcome. Typically, it involves the extremities, with less than 10 per cent occurring in the head and neck region.<sup>1</sup> It is predominant in younger adults, and males are more often affected than females. The neck and upper aerodigestive tract (oral cavity, oropharynx, hypopharynx and larynx) are the most common locations for synovial sarcoma in the head and neck region. Although it is called synovial sarcoma, it does not originate from the synovium but rather from primitive mesenchymal cells.

Synovial sarcoma involving the paranasal sinuses is a rare entity. The case reported here represents an extremely rare occurrence as it involves the ethmoidal sinus in an elderly patient. We present this case to add to the existing literature on synovial sarcoma.

### Case report

An 80-year-old woman presented at our clinic with a 6-month history of persistent right-sided epistaxis. Nasal endoscopy was performed, which identified a mass in the nasal cavity arising medial to the middle turbinate. There were no palpable lymph nodes in her neck.

Magnetic resonance imaging of the paranasal sinuses revealed a heterogeneous soft tissue lesion in the right ethmoid and sphenoid sinuses, extending into the right nasal cavity. It showed patchy gadolinium enhancement (Figure 1). The adjacent cavernous sinus, clivus and anterior cranial fossa were intact, and the brain parenchyma, ventricles and orbits were otherwise normal.

Microscopic examination of a biopsy sample from the nasal cavity mass showed that the tumour was composed of oval cells with large vesicular nuclei. There was no evidence of epithelial or glandular differentiation. Immunohistochemical staining showed that the cells were positive for vimentin,

bcl-2 and cluster of differentiation 56, and focally positive for epithelial membrane antigen. Based on the histopathological examination, it was concluded that the tumour was a monophasic synovial sarcoma. Figure 2 shows the histological findings for the haematoxylin and eosin stain, at ×400 magnification.

The patient underwent endoscopic excision of the tumour. Intra-operatively, the tumour was observed to involve the posterior septum, middle turbinate, and ethmoid and sphenoid sinuses.

The patient refused post-operative radiotherapy. Three months later, however, there was local recurrence of the tumour. The patient underwent further endoscopic excision of the tumour with post-operative radiotherapy. Unfortunately, the patient died nine months after the initial diagnosis with metastasis to the brain and neck nodes. The paranasal sinus was tumour-free.

### Discussion

Synovial sarcoma of the head and neck region typically occurs in young adults. In this case, the patient was an 80-year-old woman, well above the mean age reported in the literature.

To the best of our knowledge, only three previous cases of synovial sarcoma of the paranasal sinuses have been reported. One patient with primary ethmoid sinus synovial cell sarcoma underwent surgical resection and radiation for recurrence, but died from distant metastases nine months after the initial diagnosis.<sup>2</sup> Another patient with metastatic synovial cell sarcoma in the sphenoid sinus died two years after sinusotomy and post-operative radiation treatment.<sup>3</sup> The third report was of a 45-year-old man with maxillary synovial sarcoma who underwent Weber–Ferguson rhinotomy with total maxillectomy, and radiotherapy.<sup>4</sup>

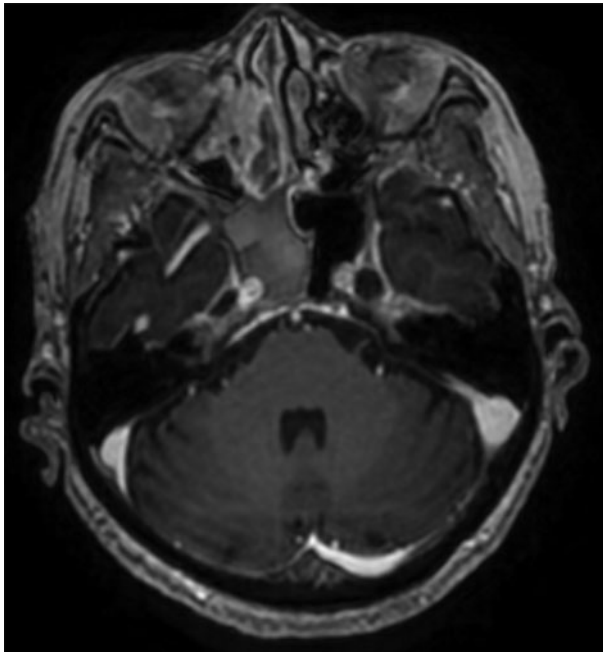


FIG. 1

Axial, post-gadolinium, T1-weighted magnetic resonance image of the patient.

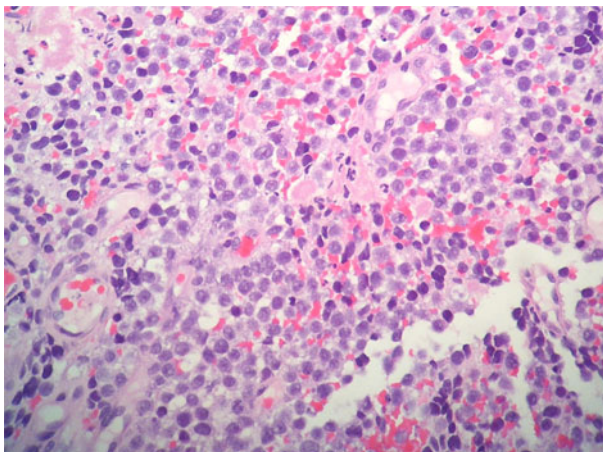


FIG. 2

Photomicrograph of the tumour showing oval cells with large vesicular nuclei. (H&E; ×400)

Microscopically, synovial sarcoma can be divided into two types: monophasic and biphasic. The fibrous monophasic type contains only spindle cells. The biphasic type contains epithelial and spindle cell components in varying proportions. The monophasic form requires confirmation of epithelial differentiation by either immunostaining or detection of cytogenetic alterations.

The accepted treatment modalities are wide excision followed by post-operative radiotherapy. Mamelle *et al.* reported that, while post-operative radiotherapy led to a decrease in local recurrence, it was not associated with any improvement in long-term survival.<sup>5</sup> In our case, the patient declined post-operative radiotherapy and there was

local recurrence of the disease three months after the first endoscopic excision.

The eventual presence of distant metastasis in our case clearly demonstrates that synovial sarcoma is an aggressive malignant tumour. This is consistent with the findings of Kartha and Bumpous.<sup>2</sup> Our patient passed away nine months after the initial diagnosis as a result of metastasis to the brain and neck nodes. The reported five-year survival rates range from 47 to 82 per cent for patients with head and neck synovial sarcoma.<sup>6,7</sup> The post-treatment recurrence rate is 50 per cent; the disease most often recurs within the first two years after treatment.<sup>8</sup>

- Synovial sarcoma of the paranasal sinuses is rare and typically affects young adults
- This report presents a case of synovial sarcoma of the ethmoidal sinus in an 80-year-old woman
- The report illustrates that synovial sarcoma can occur in elderly patients
- It adds to the existing literature on the disease progression and prognosis of this rare tumour

In conclusion, synovial sarcoma of the paranasal sinuses is rare and typically affects young adults. However, it can occur in elderly patients. It should be included in the differential diagnosis when managing tumours of the paranasal sinuses, even in the elderly. Aggressive treatment should be initiated with primary resection followed by radiotherapy, with or without chemotherapy.

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