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Synovial sarcoma of the inferior nasal turbinate: A rare case report in Sudanese patient

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ABSTRACT

Background: Synovial sarcoma is a known soft tissue malignancy that affects mainly the extremities while it is rare in the head and neck region and rarest in the inferior nasal turbinate, to our knowledge, no cases particularly in the inferior nasal turbinate have been reported to date. *Case Reports:* We managed a patient with synovial sarcoma who presented with epistaxis, right nasal obstruction and right inferior nasal turbinate mass. Endoscopic nasal biopsy and immunohistochemistry markers confirmed synovial sarcoma. As an early stage of inferior nasal turbinate synovial sarcoma, our patient was managed by endoscopic nasal surgery only. He was observed with close follow-up for 11 months and he had no signs of recurrence after his operation. *Conclusion:* Sino nasal sarcomas are rare aggressive and carry a poor prognosis. For the early stage, surgical resection is the best treatment modality. Close follow-up is warranted for the optimal outcome.

Keywords: Sino nasal sarcomas, Synovial sarcoma, malignancy, inferior nasal turbinate.

1. INTRODUCTION

Synovial sarcoma is considered a rare and high-grade soft-tissue malignant tumor accounting for 5%–10% of all adult soft-tissue sarcomas. The limbs are the most common site, but anywhere in the body may be affected (Ohan et al., 2020). It occurs in particular areas of extremities, closely associated with tendon sheath, bursae and joint capsule (Faur et al., 2021). About 3%–12% of synovial sarcomas arise in the head and neck region field (Carrillo et al., 1992; Shokralla & Fathalla, 2020). It mainly affects patients in their third decade and the male-to-female ratio is almost equal. The 5-year overall survival rates have been reported to be 44.6%–61% (Wibmer et al., 2010).

Standard treatment is complete resection, while the efficacy of adjuvant radiotherapy and chemotherapy has been studied. However, the response to chemotherapy in the head and neck region has not been reported in detail. To our knowledge, no cases of inferior turbinate synovial sarcoma have been reported globally, here; we present a case of this rare entity and discuss appropriate treatment.

2. CASE PRESENTATION

A 50-year-old Sudanese housewife was referred to our clinic with Epistaxis and right nasal obstruction for three months. The nasal obstruction was progressive and resistant to nasal decongestant and nasal steroids. Epistaxis was minimal in the amount and there were no other causes of epistaxis in the history and she did not complain of epiphora. She denied fatigue, anorexia or weight loss and she was not admitted with any medical or surgical disease in most of her adult life. In-office nasal examination revealed a significant right inferior turbinate mass which caused right nasal cavity narrowing and didn't abut on the nasal septum there was minimal ulceration on the mass and it was bleed minimally on contact.

No other masses, para nasal sinuses discharge or other ulceration was appreciated during the office endoscopy. No abnormality was detected in the opposite nasal cavity. No abnormalities were detected upon general examination and examinations of both eyes, oral cavity, brain, cranial nerves and the head and neck lymph nodes. Computed tomography showed a right nasal inferior turbinate mass occupying most of the right nasal cavity with no septal deviation and otherwise normal Para nasal sinuses (Figure 1A, 1B). The mass was only confined to the anterior part of the right inferior turbinate and the nasolacrimal duct was not obstructed.

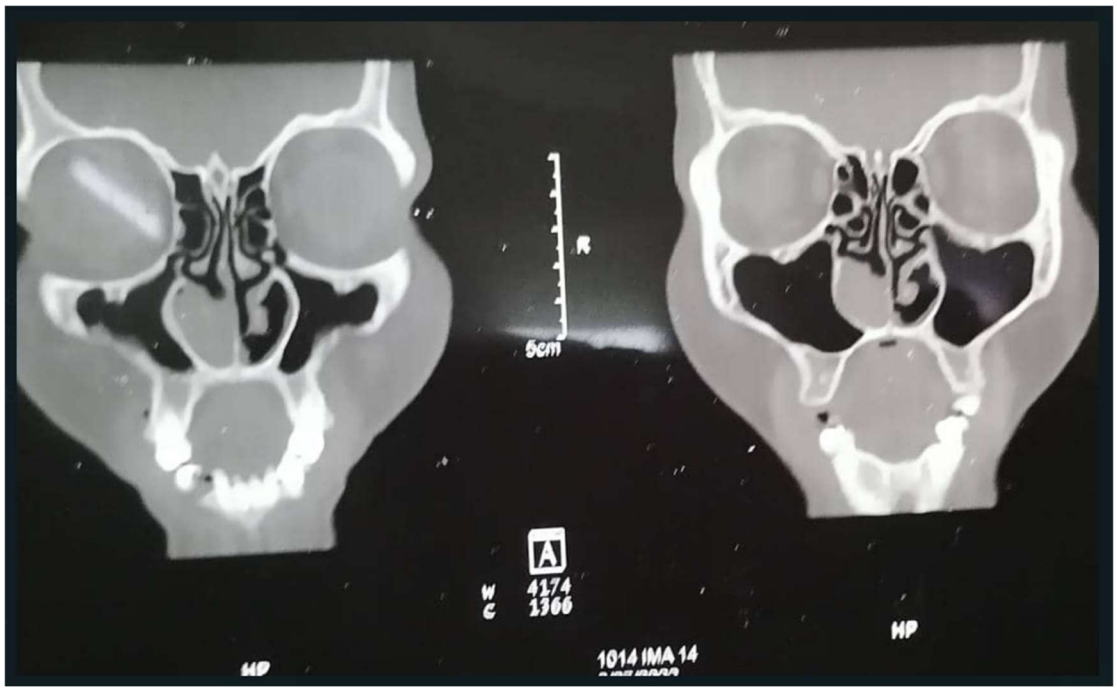

Figure 1A Computed tomography of the Sino nasal tract (coronal view) showing right inferior nasal turbinate synovial sarcoma

The decision was made to proceed with Endoscopic endonasal surgery to excise the right inferior turbinate mass for histopathological study and to improve her nasal obstruction. Endoscopic endonasal surgery in the right nasal cavity was performed and an irregular mass of the right inferior nasal turbinate with the underlying anterior part of the turbinate was surgically excised (Figure 2A, 2B). Excisional biopsy of the mass was sent as permanent specimens for pathologic review. Haemostasias secured using anterior nasal packing which was removed 48 hours following surgery. Prophylactic oral antibiotics, nasal saline drops and analgesics were prescribed postoperatively.

The gross pathology appearance of the resected mass was grey, multinodular and it was 3 cm in diameter. Histopathologic examination demonstrated right nasal inferior turbinate involvement by a high-grade sarcoma. This neoplasm was composed of monomorphic spindle cells with variable epithelial differentiation. The tissue was comprised entirely of spindle cells and showed areas of calcifications and ossification. Moreover, there were poorly differentiated areas with increased cellularity, nuclear atypia and high mitotic activity (15 mitoses per 1.7 mm2). Inflammatory cells include lymphocytes, plasma cells and foamy histocytes. The stroma was scant and showed cystic spaces and dilated blood vessels.

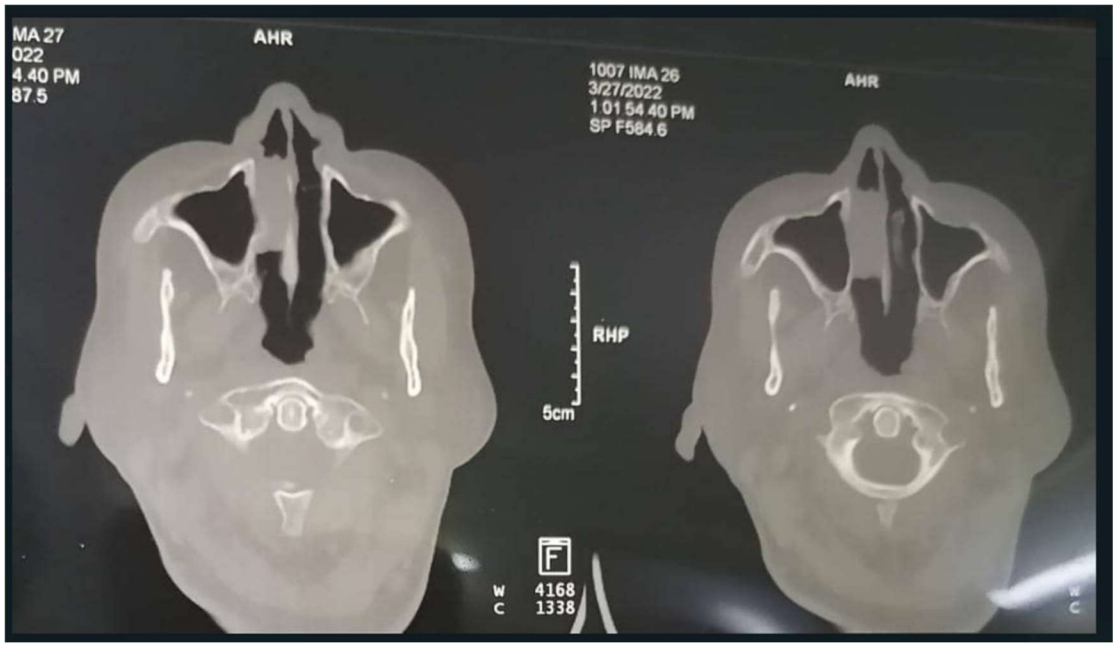

Figure 1B Computed tomography of the Sino nasal tract (Axial view) showing right inferior nasal turbinate synovial sarcoma

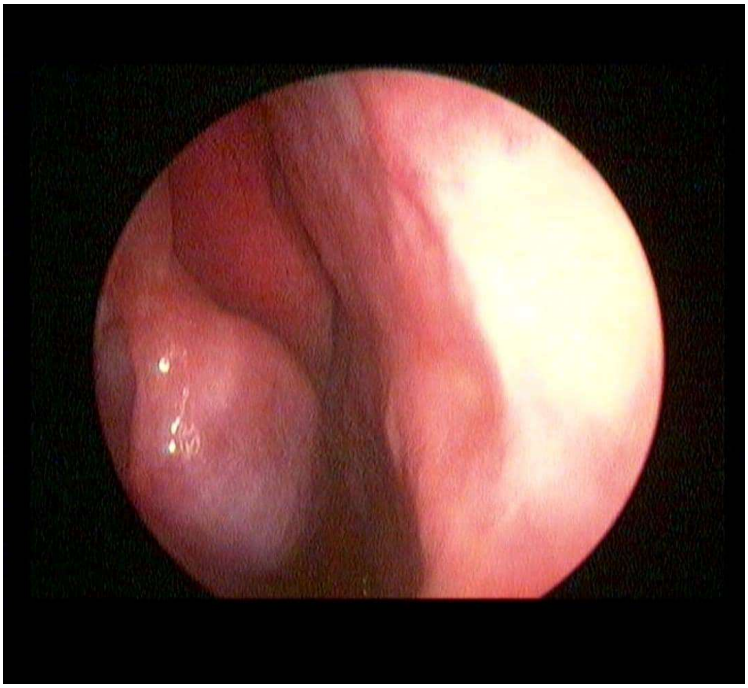

Figure 2A Post-operative inferior turbinate surgical mass reduction

On immunohistochemistry, diffuse expression of bcl-2 was seen and the tumor stain was positive for CD99. Immunohistochemistry also demonstrates strong and diffuse nuclear staining for the transcriptional corepressor and NY-ESO-1 was also expressed. Added to that immunohistochemical stains were positive for CK, while negative for S100, CD43 and SMA. A provisional diagnosis of Synovial sarcoma of the right inferior nasal turbinate was made. Subsequent PET and CT scans were negative for distal and local malignancies. The patient was observed with close follow-up and has had no signs of recurrence as of 11 months after his operation.

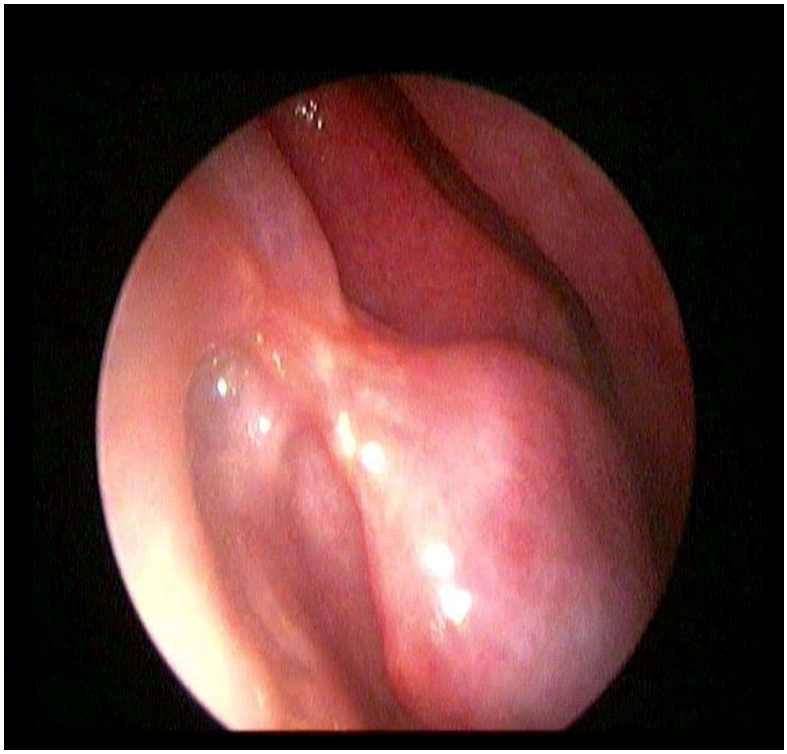

Figure 2B Post-operative inferior turbinate surgical mass reduction

3. DISCUSSION

To our knowledge, no cases of particularly inferior turbinate synovial sarcoma have been reported globally. Synovial sarcoma is known to occur in extremities while extremely rare in the head and neck, the known sites of involvement are the hypopharynx (the most common site), the oropharynx, the larynx and the soft tissues of the neck (Tos et al., 1998). Synovial sarcoma of the nose and paranasal sinuses is extremely rare, with only eleven patients having been reported in the literature (Saito et al., 2018).

Head and neck Synovial sarcoma typically present as a slowly growing mass in patients younger than 45 years. The clinical signs and symptoms according to the tumor site; may include swallowing difficulty, sore throat, hoarse voice, headaches and a palpable swelling, while in the Sino nasal tract, similar to our case, patients are known to present with nasal obstruction, epistaxis and often paranasal sinuses invasion in advanced disease field (Heslin et al., 1997). Diagnosing of head and neck synovial sarcoma is challenging and the tumor is frequently overlooked, partly because it needs a high index of suspicion.

A Computed tomography (CT) scan can indicate the malignant nature of the nasal mass; the classic CT appearance is that of a multiloculated tumor with smooth edges and heterogeneous enhancement following injection of contrast medium. Calcifications may be seen and it's typically associated with a better prognosis. When the bone invasion is suspected, a bone window CT must be requested. Magnetic resonance imaging (MRI) is crucial to determine the anatomic extent of the swelling. In our case, a Contrast type of CT scan and a diagnostic MRI were not done because they were limited in availability in Sudan at the time of the presentation.

A chest CT is useful in looking for any pulmonary metastasis and assessing the patient's medical fitness for general anesthesia. The gold standard for the diagnosis of synovial sarcoma is a tissue biopsy and histopathologic study field (Gazendam et al., 2021). The specimen obtains by an open biopsy as in our case. When the diagnosis remains uncertain despite a thorough workup another diagnostic marker might be chromosomal rearrangements. In patients who have synovial sarcoma, chromosomal alterations specifically affect the translocation between chromosomes X and 18. No chromosomal study was done in our case because it was clear from the pathology report that the diagnosis was synovial sarcoma.

Pathologic and histologic findings; the macroscopic appearance varies from cystic to solid to necrotic. Microscopically, synovial sarcoma constitutes two types: Monophasic and biphasic. The fibrous monophasic type consists of only spindled cells and the biphasic type contain epithelial and spindle-cell components in varying proportions. As our case shows, the monophasic type with criteria of a fascicular proliferation of small, uniform spindle cells. The monophasic form requires confirmation of epithelial differentiation by immunostaining or cytogenetic alterations detection. An adequate biopsy specimen is important for an accurate diagnosis.

The basic principle of synovial sarcoma surgery of resection with negative margins (R0) should be the target, although it is difficult to achieve in the Sino nasal surgery, especially endoscopic sinus surgery due to important structures close to the tumor and microdebrider/piecemeal resection, added to that, endoscopic surgery requires expertise in such cases. Despite that, the tumor was excised endoscopically since it involved only the anterior part of the inferior nasal turbinate. An open surgical approach is another option not chosen in our case since it's helpful and advantageous in advanced cases only (Ohan et al., 2020).

Synovial sarcoma in the extremities or sinonasal tract is not radiosensitive, radiation therapy of primary, recurrent or metastatic synovial sarcoma is generally ineffective in reducing tumor size. Chemotherapy for sinonasal sarcoma has only been used effectively in adjuvant with surgery and may be considered in patients with multiple metastases and/or a large primary tumor. There is a lack of literature regarding the management of the neck in patients presenting with nasal sarcomas; however, as spread tends to be locoregional, there is likely limited benefit from neck dissection or radiation to the neck.

4. CONCLUSION

Inferior nasal turbinate synovial sarcoma is a rare disease. When encountered, Endoscopic surgical excision is the preferred treatment modality, with reservation of chemotherapy for advanced disease.

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Author Contributions

Writing the manuscript: Dr Mujtaba A Ali

Reviewing the manuscript: Prof. Sharfi Ahmed

Ethical approval

The study protocol was reviewed and approved by the Omdurman Islamic University Ethical Committee with an ethical approval number (No.11/2/23).

Informed consent

Written & Oral informed consent was obtained from the participant included in the study. Additional informed consent was obtained from the participant for whom identifying information is included in this manuscript.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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