CASE REPORT

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Oropharyngeal Synovial Sarcoma: A Case Report

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ABSTRACT

Synovial sarcoma is a rare and aggressive type of sarcoma that develops from mesenchymal cells and accounts for around 8% to 10% of all cases of soft tissue sarcoma. The presence of synovial sarcoma in the head-and-neck region is characterised by aggressive behaviour and a generally unfavourable prognosis. Understanding the various manifestations of synovial sarcoma, particularly uncommon locations like the oropharynx, significantly improves patient outcomes by facilitating early diagnosis and prompt management. We report a case of a 57-year-old Malay woman who presented with a two-month history of sore throat and odynophagia, complicated with partial airway obstruction. She underwent endoscopic excision of the right oropharyngeal tumour via transoral approach, and the histopathological examination revealed monophasic synovial sarcoma. The surgical management, histopathological characteristics, and rarity of the disease are discussed.

Keywords: *Head and neck sarcoma; oropharynx; synovial sarcoma*

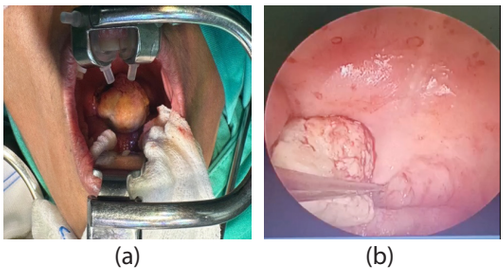
INTRODUCTION

Synovial sarcoma (SS) is a rare form of malignant tumour originating from mesenchymal precursor stem cells unrelated to mature synovial tissue. Studies in the scientific literature have emphasised the presence of this tumour in atypical locations, often far from areas lined with synovium, specifically the abdominal wall and the head and neck region (Hale & Calder, 1970; Ramamurthy *et al*., 1995). Head and neck involvement accounts for less than 10% of SS (Ramamurthy *et al*., 1995). The majority of the head and neck sarcomas are found in the cervical and hypopharyngeal region (Balakrishnan *et al*., 2012). Oropharyngeal SS is extremely rare and therefore scarcely reported in the literature.

CASE PRESENTATION

A 57-year-old Malay lady presented with a two-month history of sore throat, dysphagia, and odynophagia, which worsened upon taking solid food. However, she was still able to tolerate fluids. She experienced painless swelling in her right neck, along with a loss of weight and appetite. Otherwise, she had no difficulty in breathing, noisy breathing, cough, hoarse voice, oral ulcer, or ear pain. She also denied having spontaneous oral bleeding, halitosis, numbness, or weakness in the face and tongue.

Upon assessment, she was comfortable on room air with no stridor. There was right level II swelling measuring approximately 2 cm × 2 cm. It was firm in consistency, non-mobile, and non-tender. An intraoral examination showed a right exophytic mass with an irregular shape that rose from the right lateral wall of the oropharynx and crossed the midline, blocking part of the airway (Fig. 1a). The surface appeared ulcerated and covered with slough. The palatoglossal arch was not involved, and the uvula was midline. Otherwise, the tongue mobility was normal. A flexible nasopharyngolaryngoscopy revealed a mass in the right oropharynx that crossed the midline and obstructed the airway (Fig. 1b). The base of the tongue was not visualised. Other structures were unremarkable.



(a) (b)

Fig. 1 (a) A mass seen at the right oropharynx, irregular surface, and covered with slough; and (b) The mass was partially obstructing the airway.

A contrasted computed tomography (CT) of the neck, thorax, abdomen, and pelvis was done, which showed a large lobulated heterogeneously enhancing exophytic mass centred on the right side of the posterior oropharyngeal wall, measuring 2.1 cm × 2.8 cm × 2.9 cm, which extended medially and crossed the midline, causing significant narrowing of the oropharyngeal airway (Fig. 2). The narrowest anteroposterior (AP) diameter measures 0.3 cm at the C2 level. Laterally, the mass has poor plane with the right tonsil. Superiorly, the mass extends to the C1 level and inferiorly to the C3 level. It has a poor plane with the base of the tongue anteriorly and part of the prevertebral muscle posteriorly. There were multiple enlarged necrotic nodes at right level II, III, IV, and V, with the largest node at right level II measuring 1.7 cm × 2.3 cm. There was no distant metastasis reported.

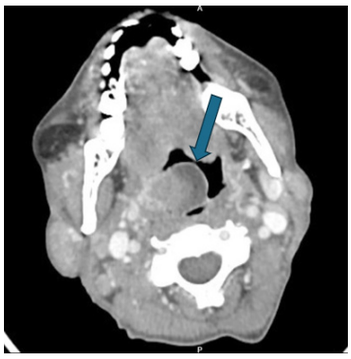


Fig. 2 A large heterogeneously enhancing mass arising from the right side of the posterior oropharyngeal wall.

Fine needle aspiration cytology of the right level II swelling was done and reported as atypia of undetermined significance. It showed a tight cluster of epithelial cells displaying oval to spindle nuclei. There were no multinucleated giant cells, necrosis or polymorphous lymphoid cells seen.

She underwent excision of the oropharyngeal mass under general anaesthesia in view of a partially obstructed airway. Endotracheal intubation was done with the aid of a C-MAC video laryngoscope. The mass was excised with an ultrasonic surgical scalpel, performed via a transoral route assisted with an endoscope (Fig. 3). The intraoperative findings revealed a pedunculated lesion arising from the lateral pharyngeal wall.

The intraoperative bleeding was minimal. Postoperatively, she recovered well, was able to resume oral feeding, and was discharged home on the next day.

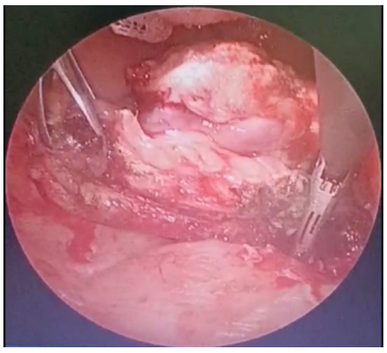
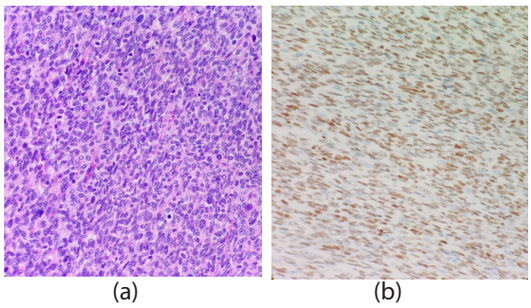


Fig. 3 Endoscopic guided transoral excision of the mass.

The histopathological diagnosis of the excised mass was consistent with monophasic SS, *Fédération Nationale des Centres de Lutte Contre le Cancer* (FNCLCC) Grade 3, composed of ovoid to spindle cells arranged in patternless sheets with vague fascicles and infiltrating the surrounding skeletal muscles (Fig. 4). One margin of resection was involved by the tumour while the other resection margin was less than 1 mm.



(a) (b)

Fig. 4 (a) Monophasic SS. Histological findings of patternless sheets of tumour cells with ovoid to spindled nuclei. (H&E with 400× magnification); and (b) Monophasic SS. Immunostain TLE1 positivity in tumour cells (400× magnification).

The patient was counselled for transoral resection of the tumour for margin clearance. However, she expressed reluctance due to her underlying comorbidities. She was then referred to the oncology team for radiotherapy and chemotherapy. Unfortunately, she also refused both treatment options, albeit after thorough discussion and counselling by the oncology team. Currently, we are monitoring her for any tumour recurrence, and a follow-up at six months post-operation revealed no recurrence (Fig. 5).

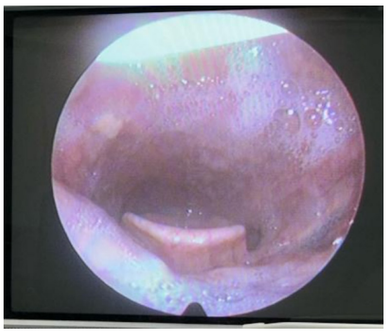


Fig. 5 During follow-up, the oropharynx was clear.

DISCUSSION

SS accounts for around 8% to 10% of all cases of soft tissue sarcoma (Ramamurthy *et al.*, 1995). Approximately 3% to 10% of sarcomas are found in the head and neck region (Balakrishnan *et al.*, 2012). Contrary to its name, SS does not originate from the synovial membranes (Siegel *et al.*, 2023). They arise from undifferentiated mesenchymal stem cells, which can differentiate into various tissue types. These tumours are named "synovial sarcomas" because of their histological appearance resembling the synovial tissue. The tumours can be found in the prevertebral space extending from the base of the skull to the hypopharynx, as well as in the retropharyngeal and parapharyngeal spaces. They also may arise from the anterior neck along the borders of the sternocleidomastoid muscle, as well as in some areas of the oropharynx and larynx (Bukachevsky *et al.*, 1992). Although it can occur in many parts of the body, it is exceptionally uncommon for it to develop in the soft palate, with just a few documented occurrences in scientific literature (Saydam *et al.*, 2002). SS exhibits a higher incidence in males (3:2 ratio) and a preference for individuals aged between 25 and 36 years (Balakrishnan *et al.*, 2012).

According to the International Classification of Diseases for Oncology (ICD-O), SS is classified into three separate histologic subtypes: monophasic, biphasic, and poorly differentiated (Fiore *et al.*, 2021). Monophasic SS, which was identified in our patient, is composed of spindle cells that are consistent in appearance. Biphasic SS is characterised by the presence of both epithelial cells organised into glandular structures and spindle cells grouped in fascicles. Poorly differentiated SS is identified by the presence of both spindle cells and round blue cells (Jayasooriya *et al.*, 2016). The two most common subtypes are monophasic and biphasic.

The FNCLCC grading system is a widely used method to evaluate the aggressiveness of soft tissue sarcomas, including synovial sarcoma (Trojani *et al.*, 1984). This system assesses three main histopathological criteria: tumour differentiation, mitotic count, and tumour necrosis. This grading system is instrumental in predicting the behaviour of sarcomas and guiding treatment decisions. Higher-grade tumours tend to be more aggressive, often necessitating more intensive treatment approaches. In this case, the FNCLCC Grade 3 indicates an aggressive tumour, necessitating a comprehensive treatment approach.

In most cases, there is a high probability of a delay in diagnosis, owing to the rarity of these tumours and the variations in their clinical manifestations. This emphasises the need to include the possibility of malignancy as the differential diagnosis of soft tissue tumours that persist or advance, especially in unusual places. The diagnosis is facilitated by imaging techniques such as CT scans and magnetic resonance imaging (MRI), which can assist in analysing the lesion, characteristics, operability, and assessing the possible existence of metastatic illness (Christie-Large *et al.*, 2008). In this case, the CT scan provided vital insights regarding the mass in the oropharynx, which played a pivotal role in planning the surgical approach.

The majority of regional and distant metastases arise due to haematogenous spread. However, up to 20% of cases had lymphatic spreads to regional lymph nodes (Bukachevsky *et al.*, 1992; Balakrishnan *et al.*, 2012). There is currently no universally accepted protocol for the treatment of SS (Bertolini *et al.*, 2003). The conventional treatment consists of the en-bloc removal of the tumour with a safe margin, while radiation plays a significant role in cases where positive or marginal margins are present (Lawrence *et al.*, 1987). Attaining safe and clear margins by resection is crucial for effective treatment, particularly in the challenging head and neck region (Bukachevsky *et al.*, 1992). The intricate structure of the head and neck region, which includes important blood vessels and nerves, sometimes makes it difficult to completely remove the tumours during the surgery. Patients who have had incomplete excision or surgical resection with tumour involvement in the surgical margin may opt for adjuvant radiation or chemotherapy (Fiore *et al.*, 2021). The use of perioperative radiation, with or without chemotherapy, may contribute to the rise in 5-year survival rates, which accounts for around 75%, seen in the last 20 to 30 years (Naing *et al.*, 2015). Prophylactic neck dissection is seldom done, with the exception when the cervical node is involved (Bukachevsky *et al.*, 1992). Following local excision, there is a significant likelihood of recurrence, often ranging from 60% to 90%, and often occurring within a period of 2 years (Italiano *et al.*, 2009; Balakrishnan *et al.*, 2012).

The usage of an ultrasonic surgical scalpel can reduce the operation time, reduce the intraoperative blood loss, and decrease postoperative pain (Tirelli *et al.*, 2014). In this case, the usage of an ultrasonic surgical scalpel was very helpful in the excision of the oropharyngeal mass, which resulted in minimal intraoperative blood loss. The patient was complaining of minimal postoperative pain and was able to resume oral feeding postoperatively. She was safely discharged home the next day.

The presence of SS in the head and neck region is characterised by its very aggressive behaviour and a generally unfavourable prognosis (Bukachevsky *et al.*, 1992; Ramamurthy *et al.*, 1995). A previous study utilising the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) programme found that the cause-specific survival rates for adults with head and neck sarcomas stood at 76%, 66%, and 61% at 2, 5, and 10 years, respectively (Peng *et al.*, 2014). While sarcomas originating outside the head and neck region predominantly result in mortality due to metastatic spread, individuals with sarcomas in the head and neck region primarily succumb to local recurrence attributed to the proximity of vital structures in the head and neck region, which can limit the possibility to attain a free resection margin (Dudhat *et al.*, 2000).

In this case, although adjuvant therapy was recommended due to the aggressive nature of the tumour and margin involvement during the surgery, the patient chose not to pursue it due to her comorbidities. She opted to focus on postoperative surveillance instead. The patient's follow-up revealed no recurrence, highlighting the effectiveness of the surgical intervention and the need for ongoing monitoring given the challenging nature of this malignancy.

CONCLUSION

This case of synovial sarcoma in the oropharynx highlights the need to address the possibility of malignancy while evaluating soft tissue tumours. Employing a multidisciplinary approach is crucial for achieving precise diagnosis and suitable treatment, as prompt identification and timely intervention significantly contribute to patient outcomes.

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