CASE REPORT

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Biphasic Synovial Sarcoma of the Cheek in a Child: A Rare Case in an Unusual Location

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ABSTRACT

Synovial sarcoma (SS) is rare in the head and neck region, accounting for only 10% of all SS, and less than 0.1% of all head and neck malignancies. Head and neck SS in paediatrics is extremely rare, with only a few cases reported worldwide. Here, we report a case of right cheek SS in a child who presented to us with a 5-month history of painless right cheek swelling. On examination, there was a palpable right cheek mass measuring 4 cm × 4 cm in size, firm in consistency, with a smooth surface and a well-defined margin. Examination of the ear, nose, throat, and head and neck was normal. Imaging showed a well-defined enhancing lesion with calcifications, with no involvement of the paranasal sinuses, intraorbital, and intracranial extension. Fine needle aspiration for cytology showed a spindle cell lesion. Excision of the right cheek tumour was done via a sublabial approach. Histopathological diagnosis of biphasic synovial sarcoma was made. Post-operatively, the patient was referred to an oncologist for adjuvant chemoradiotherapy. The patient recovered well, and no recurrence was found during the follow-up at six months.

Keywords: *Cheek; head and neck neoplasm; sublabial approach; synovial sarcoma*

INTRODUCTION

Synovial sarcomas (SS) are aggressive malignant soft tissue tumours, accounting for 5% to 10% of all soft-tissue malignancies (Herzog 2005; Harb *et al.*, 2007). SS most commonly affects the extremities, while head and neck SS remains rare, accounting for approximately 10% of all SS and less than 0.1% of all head and neck malignancies (Amble *et al.*, 1992; Sturgis & Potter, 2003). SS shows a preponderance of males in the second to third decade of life (Mettman *et al.*, 2009), and is extremely rare in paediatric groups (Lamry *et al.*, 2021).

CASE REPORT

A 9-year-old boy presented with a 5-month history of painless right cheek swelling, which was gradually increasing in size. Otherwise, there were no nasal symptoms, eye symptoms or neck swelling. He had no other comorbidities and denied constitutional symptoms. On examination, there was a palpable right cheek mass measuring 4 cm × 4 cm in size. It was firm in consistency, had a smooth surface, and a well-defined margin. Nasoendoscopy was normal with no mass or discharge from the osteomeatal complex and nasopharynx. There was no palpable neck mass or cervical lymphadenopathy. Examinations of the ears and throat were unremarkable.

Contrast computed tomography (CT) of paranasal sinuses revealed a well-defined enhancing lesion over the right cheek, measuring 2.1 cm × 2.4 cm × 2.7 cm with calcifications within. There was no erosion or remodelling of the right anterior maxillary sinus wall, nor paranasal involvement. Magnetic resonance imaging (MRI) showed T1W hypointense and T2W/STIR hyperintense right check lesion. No intraorbital or intracranial extension was seen (Fig. 1). Fine needle aspiration for cytology (FNAC) of the right cheek mass was reported as a spindle cell lesion.

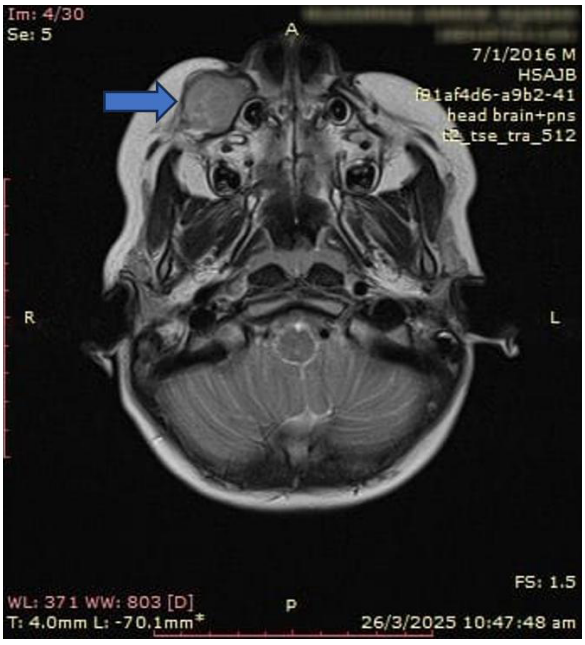

Fig. 1 MRI T1-weighted showed hypointense, well-defined right cheek lesion.

The child underwent excision of the right cheek tumour via sublabial approach, where an incision was made over the right gingivobuccal sulcus (Fig. 2). Intraoperatively, the tumour was meticulously dissected from surrounding tissues using blunt instruments. The tumour was friable upon manipulation, necessitating removal in piecemeal manner.

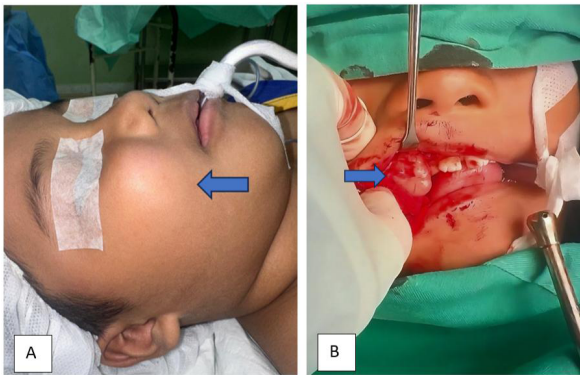

Fig. 1 (A) Preoperative image of the right cheek mass; (B) Right cheek tumour excised via sublabial incision.

Macroscopically, the tumour measured 3.0 cm × 3.0 cm × 4.0 cm, well circumscribed and unencapsulated. Microscopically, the tumour tissue is composed of neoplastic spindle cells arranged in sheets and some in fascicles. The tumour cells are enlarged, slightly pleomorphic, spindle to epitheloid in shape with vesicular nuclei, prominent nucleoli and eosinophilic cytoplasm. Mitoses and calcifications were seen. There was no necrosis area, perineural or lymphovascular invasion. The tumour cells were strongly and diffusely positive towards SS18-SSX with focal positivity towards CKAE1/AE3 and EMA (Fig. 3). A diagnosis of biphasic SS was made.

Post-operatively, the patient recovered well. He did not complain of facial numbness or blurring of vision. The wound at the gingivobuccal sulcus was well healed. He was then referred to the paediatric oncologist for adjuvant chemo-radiotherapy due to the possibility of compromised margins. Subsequent follow-up at six months showed no clinical evidence of recurrence.

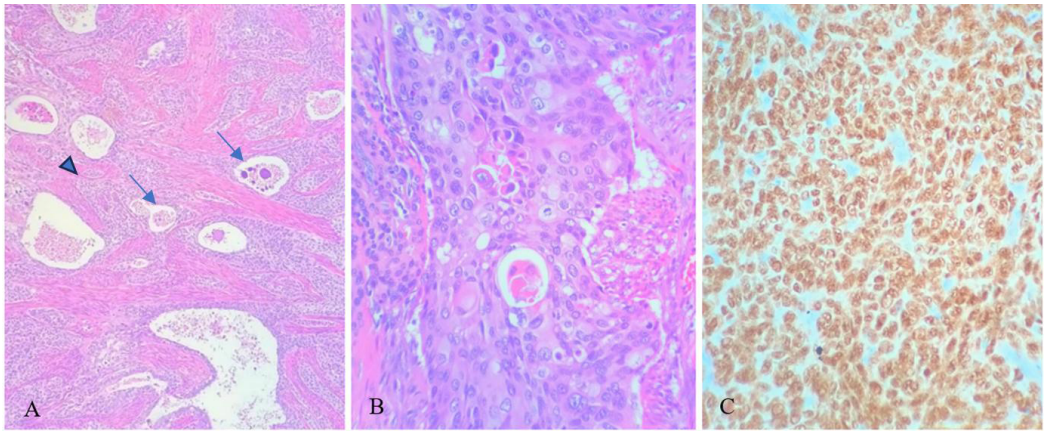

Fig. 3 Biphasic SS. (A) Scattered small blood vessels (arrows) in between neoplastic spindle cells (arrowhead); (B) Epithelial component; (C) Positive stain SS18-SSX.

DISCUSSION

Head and neck SS is rare, with the most common sites being the upper aerodigestive tract, such as the oral cavity, hypopharynx and larynx (Mettman *et al.*, 2009). Only 3% of head and neck SS occurs in the cheek (Ojha *et al.*, 2023). Head and neck SS is extremely rare in paediatrics, with only a few cases reported worldwide (Lamry *et al.*, 2021). The patients usually present with a progressive painless mass in the head and neck region (Harb *et al.*, 2007).

SS is a misnomer as it does not derive from synovium, nor expresses features of synovial differentiation. Rather, SS arises from pluripotent mesenchymal cells that undergo malignant transformation (Mettman *et al.*, 2009; Zin *et al.*, 2025). Histologically, SS is divided into three subtypes: monophasic type, biphasic type, and poorly differentiated type (Zin *et al.*, 2025). Immunohistochemically, both epithelial and spindle cells are positive for epithelial membrane antigen (EMA) and cytokeratin, while the spindle cells are positive for vimentin only (Wadhwan *et al.*, 2011). In this case, a diagnosis of biphasic synovial sarcoma was established based on the histological presence of spindle cells arranged in sheets and fascicles, alongside epithelial components that exhibit strong and diffuse positivity towards SS18-SSX.

The imaging modalities of head and neck SS include MRI and CT. The imaging of choice is MRI, which offers superior soft tissue resolution. The lesions are generally well-defined, heterogeneous, multilocular and may contain septations, punctate calcifications and haemorrhage (O'Sullivan *et al.*, 2008). CT is used to identify subtle soft tissue calcifications, bony involvement and pulmonary metastasis (O'Sullivan *et al.*, 2008).

The mainstay treatment of SS is surgical resection with negative margins followed by radiotherapy (Lee *et al.*, 2008). The surgical management of head and neck SS in paediatric groups differs from sarcoma of the extremities, in which a resection margin of 5 cm is not possible due to the proximity of vital structures in the head and neck region (Lamry *et al.*, 2021). In reported cases of large cheek SS in adults, wide local excision was done via Weber Ferguson incision in an attempt to achieve clear margins (Mettman *et al.*, 2009; Ojha *et al.*, 2023). However, in this case, taking into account the patient's young age, relatively small and well-defined tumour, with no extension into the paranasal sinus and orbit, tumour excision was planned via sublabial approach. This method also aimed to minimise postoperative complications and morbidity, particularly avoiding a noticeable facial scar, as well as the risk of nasal alar contracture, ectropion, and orbital injury.

In this case, a surgical incision was made along the right upper gingivobuccal sulcus. The tissues were dissected carefully until the tumour was adequately exposed. The tumour was meticulously separated from the surrounding tissues. However, due to its friable nature and tendency to disintegrate upon manipulation, piecemeal excision was performed. Care was taken to ensure that the tumour was removed completely. Finally, the incision was closed with absorbable suture and compression dressing was applied to the surgical site. To the best of our knowledge, this is the first reported case of cheek SS excised via sublabial approach, in contrast to previously reported cases that utilised a more extensive Weber-Fergusson incision for surgical access (Ojha *et al.*, 2023).

Postoperatively, the child was sent for adjuvant chemo-radiotherapy due to the potentially compromised tumour margin. Chemo-radiotherapy was necessary as the tumour sample sent for histopathological examination was in multiple fragments, rendering an inconclusive margin assessment. Although the dosage of radiotherapy in treating paediatric sarcoma is not standardised, non-rhabdomyosarcoma soft tissue sarcoma is usually treated with 60 Gray in 30 fractions (Lamry *et al.*, 2021). The role of chemotherapy remains controversial and is recommended as neoadjuvant therapy in tumours with more than 5 cm in size, local extension, bony involvement and higher risk surgical sites (Harb *et al.*, 2007).

The overall 5-year survival rate for SS is poor despite its relatively slow growth, and the tumour has a high recurrence rate of 60% to 90% despite therapy (Balakrishnan *et al.*, 2012; Mahesh *et al.*, 2013). The factors influencing prognosis include tumour size and extension at the time of primary treatment, both of which are inversely related (Carrillo *et al.*, 1992). Most metastases originate from hematogenous dissemination, and up to 20% spread through the lymphatics to regional lymph nodes (Bukachevsky *et al.*, 1992). Regional metastasis has been reported in 12.5% of cases of head and neck SS, while distant metastases are usually seen in the lungs and bone marrow (Ishiki *et al.*, 2009). Therefore, early diagnosis and treatment help in improving the prognosis and survival of patients. Patients should be monitored lifelong due to the tumour's tendency for late recurrence and metastasis.

CONCLUSION

Head and neck SS, especially in the cheek region, in the paediatric population, is extremely uncommon. Clinicians should maintain a high index of suspicion, as early diagnosis and timely management through a coordinated, multidisciplinary effort are essential for favorable outcomes. Surgical excision of cheek SS, if small (< 5 cm as in this case), and no locoregional extension, can be performed via sublabial approach.

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