CASE REPORT

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Alveolar Soft Part Sarcoma as a Great Masquerader of Unilateral Sinonasal Tumour in Child: A Case Report

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ABSTRACT

Alveolar soft part sarcoma (ASPS) is a rare mesenchymal neoplasm that accounts for less than 1% of all sarcoma and the most common site of tumour is found in the soft tissue of the extremity. The incidence of ASPS in paediatric age group is rare. It is extremely uncommon for the tumour arising from the sinonasal within the head and neck region. Most of the reported cases were in the orbit and tongue. Localized ASPS carries a good prognosis after complete total resection. Diagnosing ASPS can be challenging as it can be a masquerader to other sinonasal soft tissue malignancy which required the laborious process of histopathological examination, immunohistochemistry, and ultrastructural examination to solve the complexity. We encountered an 11-year-old girl who presented with sudden onset of left nasal obstruction and on examination noted a unilateral vascular mass in the left nasal cavity turned out to be ASPS. She underwent left nasal tumour debulking via endoscopic approach under general anaesthesia. Post-operative, she recovered well with regular endoscopic surveillance and was referred to the oncology team for post-operative radiotherapy.

Keywords: *Alveolar soft part; chemoradiotherapy; paediatric; paranasal sinus; sarcoma.*

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is a rare mesenchymal neoplasm that accounts for less than 0.4 to 1% of all sarcomas and is most commonly found in the soft tissue of the extremities (58%) followed by trunk (24%), head and neck region (15%) (Yigitbasi *et al.*, 2004; Tao *et al.*, 2017; Flores *et al.*, 2018). The incidence in paediatric age group is rare and the most common age group is between 15 to 35 years old (Yigitbasi *et al.*, 2004). As regards the ASPS of the head and neck, it tends to occur in younger age group of patients as compared to the ASPS of the extremity and frequently affects the region of the orbit and tongue (Yigitbasi *et al.*, 2004; Flores *et al.*, 2018).

The diagnosis of ASPS poses a huge challenge and dilemma to the clinician as it is a great mimicker to other malignancies such as renal cell carcinoma, hepatocellular carcinoma, alveolar rhabdomyosarcoma, paraganglioma, malignant melanoma, and adrenal cortical carcinoma due to their abundant of eosinophilic and clear cytoplasm (Folpe & Deyrup, 2006; Cho & Kim, 2014). With the combination of radiological, histopathological, ultrastructural, molecular genetics, immunohistochemical analysis and examination, they distinct ASPS from other types of neoplasm and soft tissue sarcoma as the management and prognosis will be greatly different (Folpe & Deyrup, 2006; Alromaih *et al.*, 2022).

CASE REPORT

An 11-year-old girl who was previously well, presented with a month history of sudden onset of left nasal block and greenish nasal discharge associated with left-sided cluster headache. She had sought treatment from the local health clinic and was prescribed a course of antibiotics without any improvement. Otherwise, she had no history of epistaxis, anosmia, shortness of breath, infective symptom, orbital symptom, toothache, loss of appetite, weight loss, dysphagia, and reduced in hearing.

On physical examination, she was normally built for her age, well hydrated, normal nutritious status, and not in respiratory distress. There was no paranasal sinus tenderness, no infraorbital numbness, no proptosis, no facial swelling, no intraoral mass, no loose tooth, and no palpable cervical lymph node. The anterior rhinoscopy was unremarkable except for the bilateral inferior turbinate hypertrophy and the cold spatula test showed reduced misting over the left side. Rigid nasoendoscopy over the left nostrils revealed a reddish mass occupying the left posterior half of the left nasal cavity, firm in consistency, friable in nature, bled upon probing, insensate, unable to pass through the scope till the nasopharynx and unable to identify the origin of the mass (Fig. 1). Otherwise, nasoendoscopy through the right nostril was clear till the nasopharynx. Immediate biopsy was done under local anaesthesia uneventfully as the mass was suspicious of malignancy based on the clinical presentation and nasoendoscopy findings.

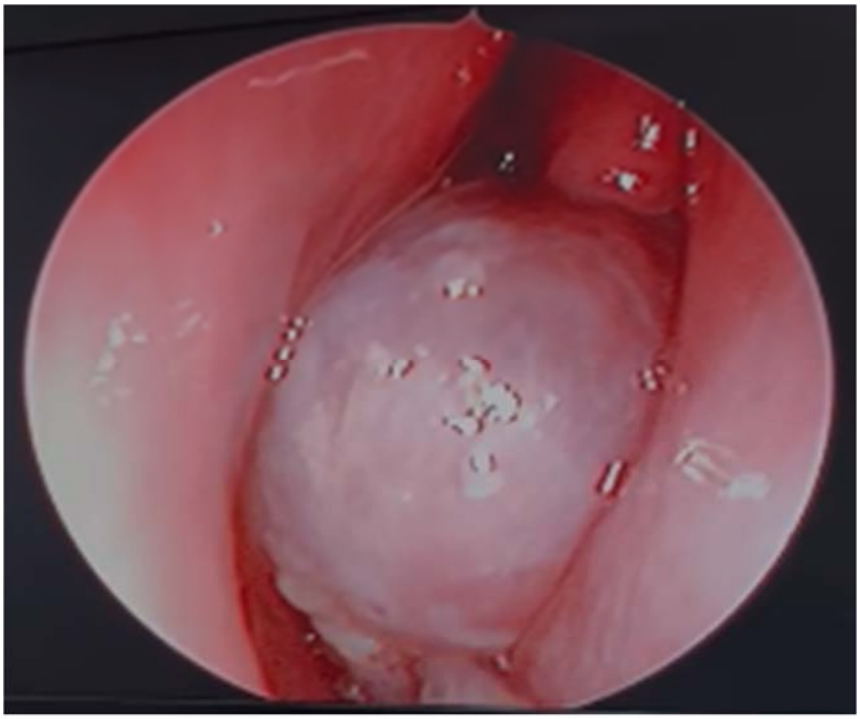

Fig. 1 Preoperative endoscopic view of the left nasal cavity showing a reddish friable mass occupying the posterior half of the left nasal cavity and unable to pass the scope through to the nasopharynx.

As expected, the histopathological examination turned out to be a malignant lesion, identified as ASPS. Urgent computed tomography of the paranasal sinus and full staging from the neck to the abdomen revealed heterogenous enhancing mass in the left nasal cavity with mass effect on the nasal septum and lateral nasal wall (Fig. 2). Besides that, the mass had extended to the left sphenoid sinus, left frontal recess, and left pterygopalatine fossa. Otherwise, there was neither intracranial or intra-orbital extension nor evidence of distant metastases.

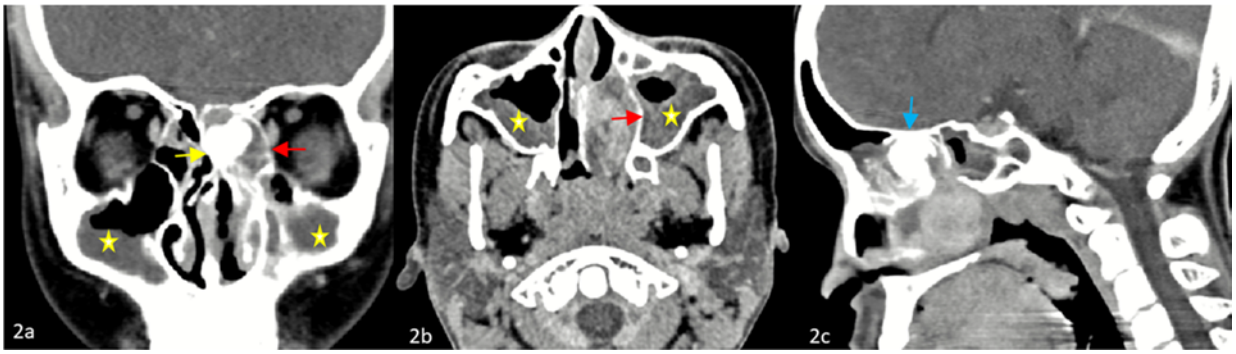

Fig. 2 Computed tomography of paranasal sinus in coronal view (a), axial view (b), and sagittal view (c). The left nasal mass has resulted bowing of the left lamina papyracea (red arrow) as well as the left nasal septum (yellow arrow). Fortunately, there is no breaching of mass to the base of skull (blue arrow). Note that there is an increase in attenuation at bilateral maxillary sinuses signifying mucous retention.

She underwent the endoscopic left nasal tumour excision and biopsy under general anaesthesia after her parents consented. Intraoperatively, the mass was noted to extend to the left lamina papyracea laterally, abutting the skull base superiorly and the left nasal septum medially (Fig. 3). Macroscopically, the mass was managed to be removed completely without any major complication or iatrogenic injury to the adjacent structures.

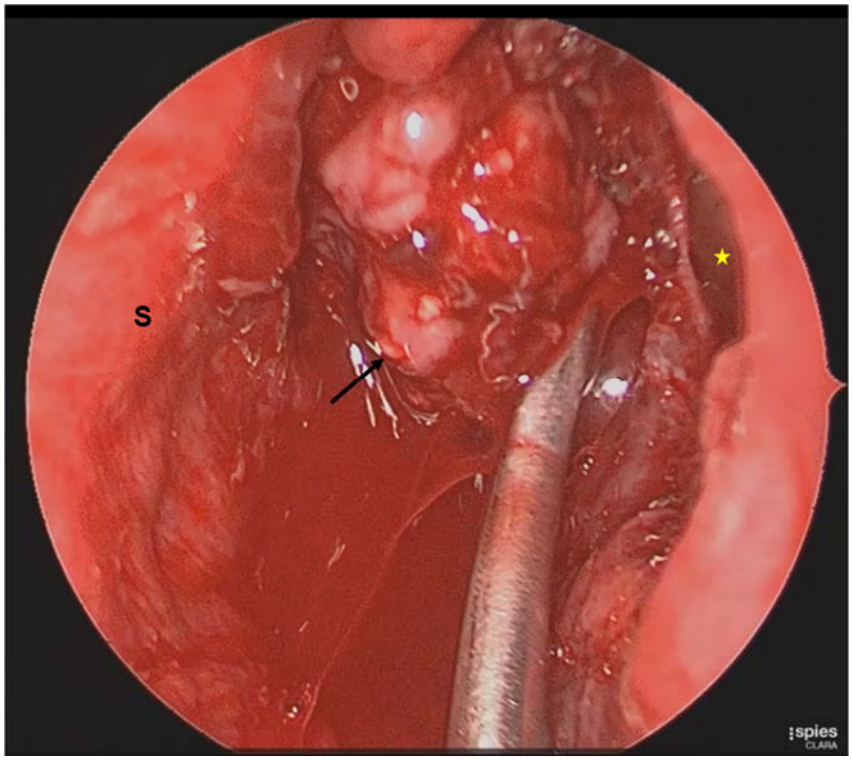

Fig. 3 Intraoperative endoscopic view of the left nasal cavity with the black arrow showing the main tumour bulk. Yellow star = left maxillary sinus ostium, S= Nasal septum.

The patient was discharged well on day 2 post-surgery after the removal of Merocel nasal packing and she was given a 1-week follow-up appointment. The final reported histopathological examination revealed similar findings of ASPS based on the microscopic appearance and immunohistochemistry-stained features (Fig. 4). Four months after surgery, patient was doing well without signs of recurrence during the surveillance nasoendoscopy examination. Repeated paranasal imaging with computed tomography and magnetic resonance imaging (MRI) revealed mucosal thickening over the left posterior nasal cavity, left ethmoidal sinus, and left sphenoid sinus suggestive of post-operative inflammatory changes without evidence of residual or relapse of the disease. She was planned for MRI surveillance over the brain and paranasal sinus every 6 month as well as monthly nasoendoscopy surveillance. She was referred to paediatric oncologist but was not started with radiotherapy in view of no evidence of relapsing of disease.

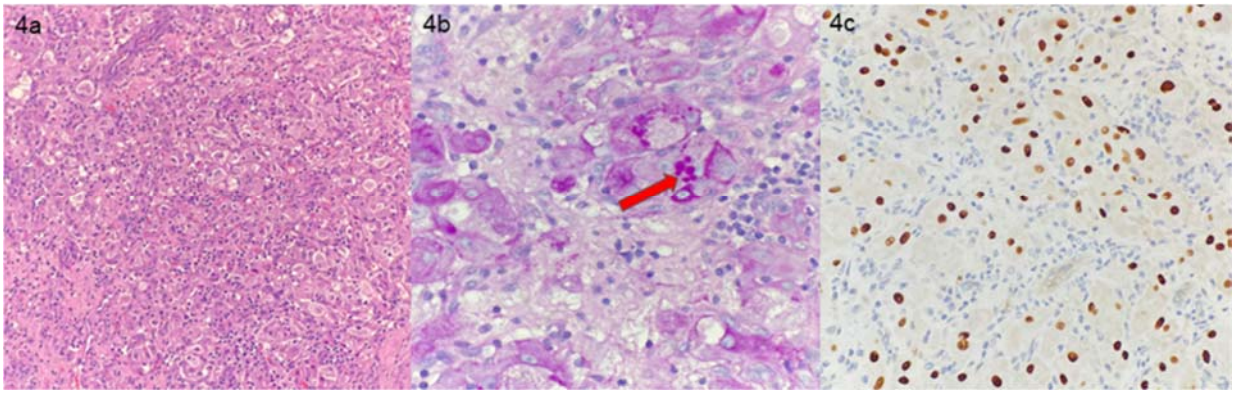

Fig. 4 (a) Microscopic examination (hematoxylin-eosin stained) revealed polygonal-shaped tumour cells with well-defined border displaying eosinophilic granular to clear cytoplasm accompanied by rounded central vesicular nucleus and prominent nucleoli. These tumour cells arranged in nests, clusters and some in organoid pattern (original magnification x400). (b) Presence of periodic acid-Schiff test positive and diastase resistant intracytoplasmic granules (red arrow) suggestive of ASPS (original magnification x600). (c) Immunohistochemistry test showed the tumour cells were positive with transcription factor E3 stained (original magnification x400).

DISCUSSION

Although ASPS manifests as a rare and slow-growing tumour, it is of utmost importance to establish the diagnosis promptly due to its tendency to metastasize. Unlike the majority of soft tissue sarcomas, ASPS frequently metastasizes early, primarily to the lungs (in 42% of cases), bones (19%), brain (15%), and lymph nodes (7%), and it has a high recurrence rate following conservative surgical excision (Tao *et al.*, 2017). As the ASPS is an indolent tumour, most of the patients were asymptomatic and came late with a high incidence of metastasis rate by the time of diagnosis (Orbach *et al.*, 2013; Cho & Kim, 2014; Jaber & Kirby, 2015). Despite evidence of metastasis, prior large case series showed prolonged survival in ASPS as compared to other soft tissue sarcoma (Lazar *et al.*, 2007; Hagerty *et al.*, 2020). In contrast, our patient reported here had an early onset of symptom within one-month possibly due to the location of the tumour obstructing the left nasal passage and reduced nasal airflow. These had caused great discomfort to the patient and significantly affecting the patient's quality of life. Fortunately, she sought medical attention swiftly which contributed to a better prognosis before the distant metastasis took place.

Factors such as tumour size of more than 5 cm, distant metastasis at the time of diagnosis, older age group more than 17 years old, male gender, and truncal primary site are associated with poor prognosis and lower overall survival rate (Wang *et al.*, 2016; Flores *et al.*, 2018). Conversely, patient has a longer survival if the tumour size is less than 5 cm, without evidence of distant metastases and clear tumour resection margin (Cho & Kim, 2014). This patient reported here had all the favourable factors as described earlier due to the small localised nasal tumour which was amenable for clear tumour resection macroscopically, younger age group, female and no evidence of distant metastasis at the time of diagnosis.

Up till today, there is still no consensus on the best treatment options for the patients. Most of the authors suggested radical surgical excision with tumour-free margin (Portera *et al.*, 2001; Cho & Kim, 2014). However, adjuvant radiotherapy can be added for the prevention of local recurrence when surgical margin involvement is suspected (Cho & Kim, 2014). Radiotherapy has been suggested for those patients with a high risk of tumour recurrence based on the clinico-

pathological evidence positive of surgical margin microscopically, tumour size, anatomic location, and local recurrence (Portera *et al.*, 2001; Kumar *et al.*, 2019). Previous case series reported combination of surgery and radiotherapy results in better overall survival in locoregional disease compared to surgery alone (Wang *et al.*, 2016). In our case, the ASPS was encountered in the nose and paranasal sinus region and technically it was difficult to achieve clearance of tumour margin microscopically due to the nature of the anatomy which is adjacent to vital structures such as the orbits, pterygopalatine fossa, skull base and intracranial space. However, the paediatric oncologist decided not to commence on post-operative radiotherapy in our patient as the repeated MRI brain showed no evidence of recurrence. Moreover, the risk of radiotherapy toxicity and potential side effect to the surrounding vital organs outweigh its benefit such as osteoradionecrosis of craniofacial bone, cataract, spinal cord, retinal and optic nerve damage.

The role of conventional chemotherapy is disappointing due to its relatively chemo-insensitivity and ineffective in ASPS, with the response rate ranging from 6%-17% (Reichardt *et al.*, 2003; Orbach *et al.*, 2013; Wang *et al.*, 2016). Furthermore, studies have shown that multimodal therapy did not improve the overall survival rate (Hagerty *et al.*, 2020; Fujiwara *et al.*, 2022). Despite the disappointing result of conventional chemotherapy, newer targeted therapy aiming to prolong the survival rate of patients is being investigated and the result is promising (Paoluzzi & Maki, 2019).

In view of ASPS is highly vascular, antiangiogenic agent and immune stimulating therapy targeting the vascular endothelial growth factor (VEGF) and hepatocyte growth factor receptors (HGFR) such as sunitinib, cediranib, sorafenib, pazopanib, or bevacizumab have been proposed (Fujiwara *et al.*, 2022). A clinical trial on 34 patients with metastatic ASPS showed an overall response rate of 35% (Kummar *et al.*, 2013). The survival period was prolonged for those patients with metastatic ASPS who received pazopanib as compared to conventional cytotoxic doxorubicin (Fujiwara *et al.*, 2022). Moreover, sunitinib in advance ASPS revealed 55% of partial response rate (Stacchiotti *et al.*, 2011). Nevertheless, studies had found that surgical resection did prolong patient survival even in stage IV metastatic disease. Systemic targeted therapy is only reserved for tumour recurrence, inoperable tumour and to facilitate tumour shrinkage before surgery (Hagerty *et al.*, 2020). Therefore, this patient was not indicated for any systemic targeted therapy due to amenability of clear tumour resection and no evidence of recurrence thus far.

Despite stable disease of ASPS post-treatment in this patient, she should be on long-term regular follow-up due to the risk of metastasis. There were evidence of progression of disease and metastasis even after a very long interval of between 99 to 252 months due to the indolent nature of ASPS (Reichardt *et al.*, 2003).

CONCLUSION

Sinonasal ASPS is a rare mesenchymal neoplasm particularly in the paediatric population and it can mimic other sinonasal soft tissue malignancies. Despite the advancement of targeted therapy, aggressive radical resection with clear tumour margin is still the preferred option of treatment when feasible. Radiotherapy can be commenced for inoperable cases or when the tumour margin is questionable. Long-term follow-up with surveillance nasoendoscopy is essential due to the risk of local recurrence. Due to the scarcity of cases, future case review in regards to the sinonasal ASPS is warranted to provide the best treatment options for the patient.

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