Malaysian J Pathol 2015; 37(1) : 53 – 56

CASE REPORT

Sinonasal haemangiopericytoma: histomorphology and differential diagnoses

Wei Qiang LEOW and Ivy SNG

Department of Pathology, Singapore General Hospital

Abstract

A 39-year-old female presented with a fleshy nasal polyp occluding the left nasal cavity, associated with haemopurulent discharge. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) scans of the paranasal cavities revealed a large polypoid tumour arising from the left middle turbinate and obstructing the left maxillary sinus ostium. However, no bony or intracranial involvement was identified. A biopsy revealed a tumour with small blue round cell morphology. The tumour cells showed diffuse strong membranous CD99 positivity and patchy CD34 positivity. Ancillary cytogenetic tests for the EWSR1 and SS18/SYT gene translocations were negative. In view of the non-invasive nature of the tumour and the low cell proliferative index (Ki-67) of 5%, a medial maxillectomy resection was performed. The resection revealed additional areas with spindle-cell morphology and focal haemangiopericytic vasculature. The tumour continued to show immunoreactivity to CD99 and CD34, as well as Smooth Muscle Actin (SMA) and Muscle Specific Actin (MSA). The overall findings are in keeping with a sinonasal haemangiopericytoma. With clear surgical resection margins, the patient is on routine follow-up and is currently disease-free.

Key words: nasal tumour, haemangiopericytoma, CD99, CD34

INTRODUCTION

Sinonasal hamangiopericytoma (HPC) is an uncommon tumour of the upper aerodigestive tract. Common presenting symptoms include epistaxis and nasal obstruction. Histologically, sinonasal HPCs are composed of a monotonous population of cells with spindled-to-round nuclei and eosinophilic-to-clear cytoplasm, arranged in fascicles and sheets. More strikingly, there is an accompanying haemangiopericytic blood vasculature, composed of staghorn-like blood vessels with perivascular hyalinization. We discuss the features of this tumour and important differentials in its diagnosis.

CASE REPORT

A 39-year-old female of Indian descent presented with a fleshy nasal polyp occluding the left nasal cavity, associated with haemopurulent discharge of six months duration. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) scans of the paranasal cavities revealed a large polypoid tumour arising from the left middle turbinate and obstructing the left maxillary sinus ostium. However, no bony or intracranial involvement was identified. An initial biopsy revealed a cellular tumour with an uniform small blue round cell morphology. As the tumour cells showed diffuse strong membranous CD99 positivity, while being negative for BCL-2, desmin and CD45; a preliminary diagnosis of Ewing's sarcoma was considered. However ancillary cytogenetic tests for EWSR1 gene translocation were negative. The patient subsequently underwent a left endoscopic medial maxillectomy resection. Intraoperatively, a left nasal polyp was seen, originating from the lateral nasal wall from the anterior ethmoidal region and extending to the maxillary ostium.

Pathology

Grossly, multiple fragments of tan beige tissue were received. Microscopically, the tumour showed a cellular proliferation of predominantly spindled and focally rounded cells, forming short fascicles and accompanied by focal staghorn-like vascular spaces. The tumour cells had

Address for correspondence: Dr Leow Wei Qiang, Pathology Department, Singapore General Hospital, 20 College Road, Academia, Level 10, Diagnostics Tower, Singapore 169856. Email: leow.wei.qiang@sgh.com.sg

scant eosinophilic to clear cytoplasm, vesicular nuclei and indistinct nucleoli (Fig. 1). Up to 10 mitoses per 10 high power fields were noted, with a Ki-67 cell proliferative index of 10%. Immunohistochemistry revealed that the tumour cells showed diffuse vimentin and CD99 positivity. There was patchy positivity for Smooth Muscle Actin (SMA), Muscle Specific Actin (MSA) and CD34 (Fig. 2). The tumour cells were negative for BCL-2, EMA, desmin, CD45 and S100.

Differential diagnoses

As the initial biopsy specimen showed a small blue round cell appearance, we considered these main differential diagnoses: Ewing's sarcoma, synovial sarcoma, rhabdomyosarcoma and lymphoma. When the subsequent resection specimen showed additional areas of spindled cells and focal haemangiopericytic vasculature, the diagnosis of sinonasal HPC was considered (Fig. 1). Immunohistochemistry with SMA and MSA confirmed the pericytic myoid nature of the tumour cells. In addition, repeated cytogenetic testing for EWSR1 and SS18/SYT gen translocations were negative, ruling out Ewing's sarcoma and synovial sarcoma. With clear surgical resection margins, the patient is on routine follow-up and is currently disease-free.

DISCUSSION

Sinonasal HPC is an uncommon sinonasal neoplasm, commonly presenting with epistaxis and airway obstruction, with symptoms averaging 10 months in duration.[1] It is an indolent neoplasm with an excellent prognosis after complete surgical resection, demonstrated by a 5-year survival rate greater than 90%. [1,2] Although local recurrence has been reported in 7% to 40% of cases, this is mostly in part due to incomplete surgical resection.[1] Cases with aggressive behaviour are rare and are usually large tumors (>5 cm), show bone invasion, have profound nuclear pleomorphism, necrosis and have high mitotic (4 mitoses /10 high-power fields) and cell proliferation (10% by Ki-67) rates.[1]

Histologically, sinonasal HPC is a cellular neoplasm composed of monotonous cells containing spindled-to-round nuclei with vesicular-to-hyperchromatic chromatin and eosinophilic-to-clear cytoplasm with indistinct cell borders.[1] Mitotic figures are generally inconspicuous and necrosis is absent. In addition, there are characteristic staghorn-like blood

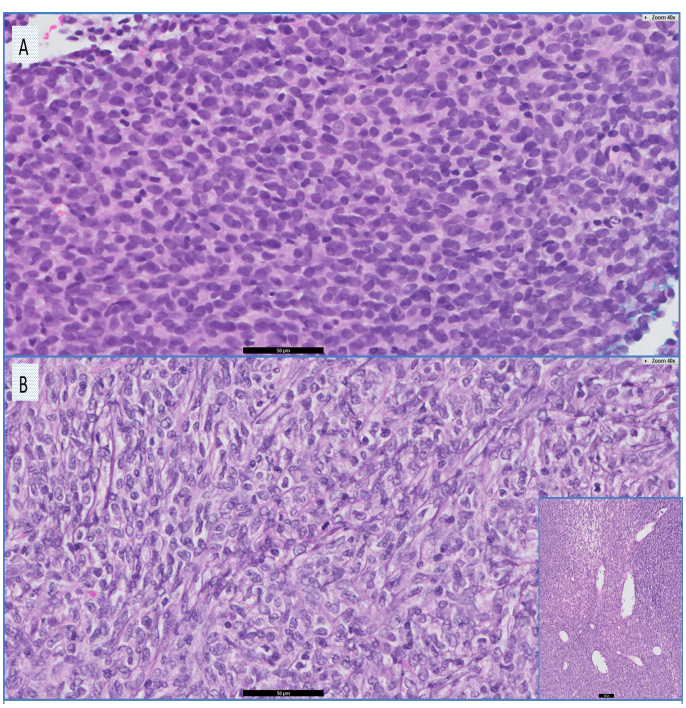

Fig. 1 (A) Initial biopsy with small blue round cell morphology (H&E, original magnification x400). (B) Resection specimen with spindled cell morphology (H&E, original magnification x400). Insert shows focal haemangiopericytic vasculature (H&E, original magnification x100).

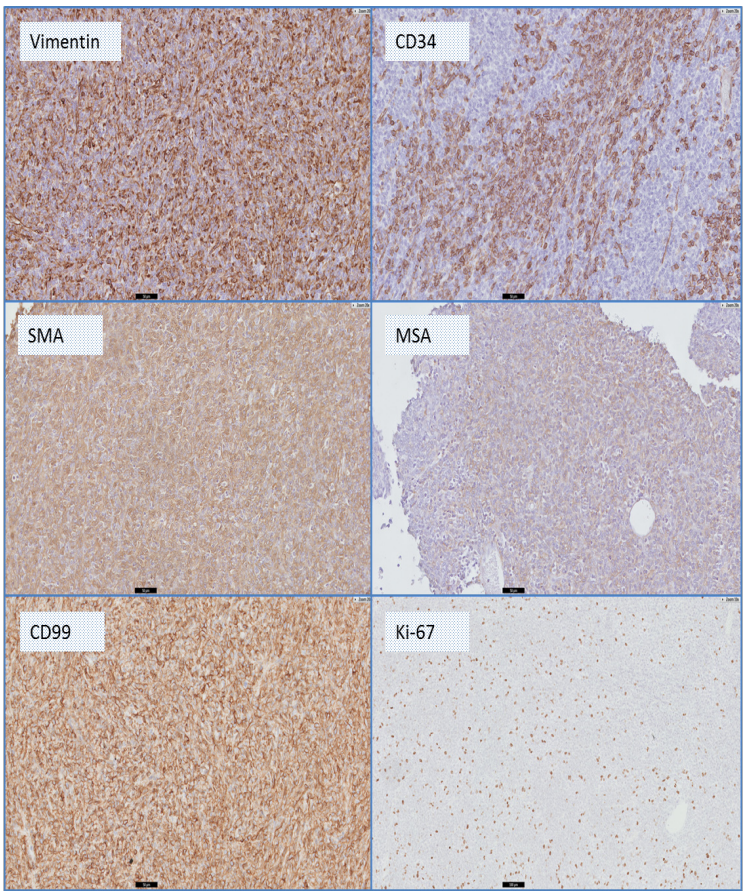

Fig. 2 Immunohistochemical stains revealing the myoid differentiation of the tumour with diffuse vimentin and SMA positivity. There is patchy MSA and CD34 positivity. Ki-67 shows the cell proliferative index to be approximately 10%. Of note, the tumour in our cases showed diffuse membranous CD99 positivity (Immunohistochemistry, original magnification x100 to x200).

vessels and frequent perivascular hyalinization. An associated inflammatory cell infiltrate and extravasated red blood cells was noted in the majority of cases.[1,2]

In our case, the initial biopsy showed a uniform small blue round cell morphology and hardly any spindled cells were discerned. Also, there were no staghorn-like blood vessels. As such, sinonasal HPC was not considered in the initial differential diagnoses. We would like to highlight this potential pitfall as sinonasal HPC may only show patchy areas of spindled cells and haemangiopericytic vasculature. It is important to have a high index of suspicion and perform ancillary tests to look for any pericytic myoid differentiation.

Some other interesting features that have been reported include a resemblance to glomus tumour, solitary fibrous tumour with keloid-like collagen deposition, lipomatous change, extramedullary hematopoiesis and multinucleated giant cells.[1,2]

Sinonasal HPCs are true pericytic tumors that exhibit a myoid phenotype demonstrable with immunohistochemistry and electron microscopy.[3] Hence such neoplasms typically stain diffusely positive for vimentin. They are also positive for SMA and MSA, although sometimes in a patchy fashion.[1,2] Staining for desmin, CD34 and S100 is typically negative, although staining for CD34 and S100 can be focally and weakly positive in a small percentage of tumours.[1,2] Additional negative stains include BCL-2, CD99, CD117, cytokeratins and caldesmon.[1] In our case, we reported diffuse strong membranous CD99 positivity in both biopsy and resection specimens. In our literature search, CD99 has never been reported as positive in a sinonasal HPC. In addition, SMA and MSA staining was patchy in our case and may easily be interpreted

as negative in small biopsies. These factors may lead the interpreting pathologist to consider Ewing's sarcoma as a differential. However we found cytogenetic testing for EWSR1 gene translocation to be a useful tool in excluding this differential.

Ultrastructural examination reveals well-developed actin thin filaments and subplasmalemmal plaques. These features suggest true pericytic differentiation of such tumors. We see electron microscopy as another useful ancillary tool in confirming the pericytic nature of the tumour cells.

ACKNOWLEDGEMENT

This material was presented as a poster during the Singhealth-Duke NUS Scientific Congress 2014. The authors have no conflicts of interest to disclose.

REFERENCES

1. Thompson LD, Miettinen M, Wenig BM. Sinonasal-type hemangiopericytoma: a clinicopathologic and immunophenotypic analysis of 104 cases showing perivascular myoid differentiation. Am J Surg Pathol. 2003; 27(6): 737-49.
2. Dandekar M, McHugh JB. Sinonasal glomangiopericytoma: case report with emphasis on the differential diagnosis. Arch Pathol Lab Med. 2010; 134(10): 1444–9.
3. Watanabe K, Saito A, Suzuki M, Yamanobe S, Suzuki T. True hemangiopericytoma of the nasal cavity. Immunohistochemical and electron microscopic study of 2 cases and a review of the literature on sinonasal hemangiopericytomas. Arch Pathol Lab Med. 2001; 125: 686-90.