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Case Report

A Case Report of Sinonasal Hemangiopericytoma

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Sinonasal hemangiopericytoma is a rare sinonasal mesenchymal tumor, and is considered different from other reported hemangiopericytomas. A 37-year-old man presented with intermittent left epistaxis for three days. Rhinoscopic examination revealed a smooth red mass in the left middle nasal meatus. A pathology report of the biopsy specimen showed sinonasal hemangiopericytoma. Magnetic resonance imaging demonstrated the tumor had eroded the posterior nasal septum, involved the bilateral posterior ethmoid sinuses, and obstructed the ostium of the left maxillary sinus causing left maxillary sinusitis. After transarterial embolization, the tumor was endoscopically excised with a microdebrider. The postoperative course was uneventful and epistaxis did not recur during the following two years. Follow-up magnetic resonance imaging did not show any tumor recurrence, although the bilateral maxillary sinusitis remained. Nevertheless, the patient should be followed regularly to exclude the possibility of recurrence or metastasis.

Key words: hemangiopericytoma, sinonasal neoplasm, hemangioma, epistaxis, pan-sinusitis

Introduction

Hemangiopericytoma is a rare (< 1%) hemangioma developing from the pericytes of Zimmerman[1]. Fifteen to thirty percent of all hemangiopericytomas occur in the head and neck, and in this region, hemangiopericytomas are predilected to the sinonasal region[1,2]. Sinonasal hemangiopericytoma (SNHP) are rare sinonasal mesenchymal tumors; however, they are considered different from other reported hemangiopericytomas[2,3]. They are also called hemangiopericytoma-like tumors, which means that they have the same origin as hemangiopericytomas, but their pathologic and clinical behavioral patterns are not necessarily the same[1,4,5]. SNHP most often present with unilateral nasal obstruction and/or epistaxis, with a polypoid mass in the nasal cavity[2,4]. Only one case has been found in a new regional hospital, opened in July 2008, in northern Taiwan. We report this rare case herein.

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Case Report

A 37-year-old man, who had a history of hypertension and had taken regular antihypertensive medication for at least five years, presented with intermittent left epistaxis for three days. Nasal packing at a private clinic stopped the epistaxis, but fresh blood leaked around the

nasal packing after light exercise at home. His blood pressure was 139/91 mmHg with a heart rate of 80/min. A physical examination revealed a smooth red mass without touch bleeding in the left middle nasal meatus (Fig. 1A). After a local biopsy was performed, pathology reported the tumor to be composed of ovoid to spindled cells arranged in intricate vasculatures and the interior of the vasculature to be dilated with staghorn-like vascular spaces (Fig. 1B). The tumor manifested with smooth-muscle actin, vimentin, CD-34 and Ki-67, rather than S-100 protein. In addition, the proliferative index was 7%, and there were occasional mitoses without any necrosis or pleomorphism, confirming the diagnosis of SNHP.

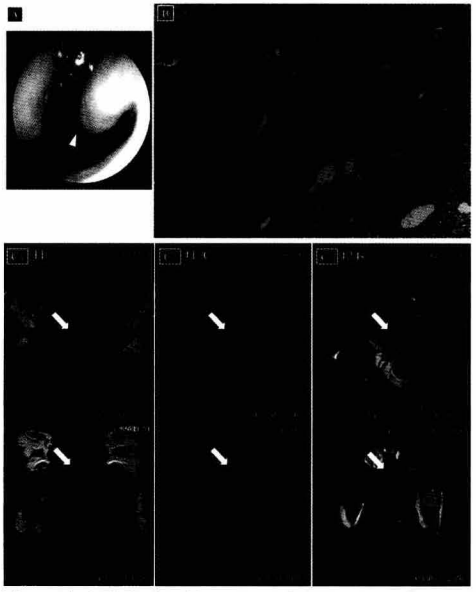

Figure 1. A: Anterior rhinoscopy shows a smooth mass in the left middle nasal meatus (arrow-head). B: Pathology shows the tumor is composed of ovoid to spindled cells arranged in intricate vasculatures. The interior of the vasculature is dilated by staghorn-like vascular spaces (filled arrow) (H&E, 200×). C: Magnetic resonance imaging shows of the tumor showed intermediate signal intensity under T1-weighted and T2-weighted fat-saturation imaging (T2 fs), and good enhancement on gadolinium-enhanced T1-weighted imaging (T1+C) (hollow arrows).

Magnetic resonance imaging (MRI) demonstrated the tumor had eroded the nasal septum, involved the bilateral posterior ethmoid sinuses and obstructed the ostium of the left maxillary sinus causing left maxillary sinusitis. Furthermore, the bilateral sphenoid and right maxillary sinuses also showed mucosal thickening and fluid collection, compatible with pan-sinusitis (Fig. 1C). Surgery was performed to prevent continued epistaxis.

Presurgical transarterial embolization was performed with 3.0 mL trisacryl microspheres (Embosphere® 500~700 um, BioSphere Medical, France) with a Fr. 2.7 micro-catheter system (Progreat TM, Terumo, Japan) placed in the left internal maxillary artery (Fig. 2). Then, the patient received general anesthesia and endotracheal intubation. Under sinonasal endoscopy, the tumor, posterior nasal septum, and the bilateral posterior ethmoid sinuses were excised with a microdebrider, and left middle meatal antrostomy was performed. The surgical margin was pathologically free of

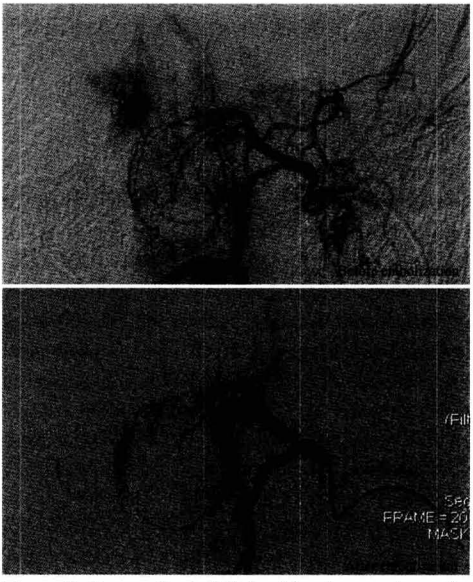

Figure 2. Concomitant digital subtraction angiography during presurgical transarterial embolization of the left internal maxillary artery (dotted circle).

tumor cells. After nasal packing for one day, the patient was discharged without any complications. The postoperative course was uneventful and epistaxis did not recur over the following two years. Follow-up magnetic resonance imaging did not show any tumor recurrence although the bilateral maxillary sinusitis remained.

Discussion

Epistaxis is a common condition in outpatient and emergency medicine, and can be caused by trauma (surgery), hypertension, septal deviation, allergic rhinitis or neoplasms[6]. Nasopharyngeal carcinoma is one of the most common attributable neoplasms, but SNHP is quite rare. SNHP is also called sinonasal-type hemangiopericytoma[2,3], hemangiopericytoma-like intranasal tumor and sinonasal hemangiopericytoma-like tumor[1]. It usually presents in the sixth and seventh decades in both genders[2,4]. Our case patient was much younger than most patients.

Contrast-enhanced computed tomography can reveal the mass effect of SNHP distorting or destroying the adjacent bony structure more clearly than plain sinus radiographs; however, gadolinium-enhanced MRI is the best tool for evaluating its vascularization and extension, and differentiating it from obstructive sinusitis. SNHP appears isointense on T1-weighted imaging, diffusely enhanced after gadolinium enhancement, and isointense to hypointense on T2-weighted imaging, as in our case (Fig. 1C). However, on post-gadolinium MRI, the inflammatory mucosa adjacent to the neoplasm was enhanced, mimicking a possibly larger neoplasm.

Histologically, SNHP contains ovoid or spindled cells with diffuse growth and prominent perivascular hyalinization, but mitosis or necrosis is seldom found. The interior of the vasculature is dilated with staghorn-like vascular spaces. The cells stain for vimentin (98%) and smooth-muscle actin (92%), factor XIIIa (78%), muscle-specific actin (77%), laminin (52%), Ki-67 (48%), CD 34 (8%), S-100 protein (3%), CD68 (2%), glial fibrillary acidic protein (2%), and bcl-2 (2%); they do not stain for epithelial, melanocytic or neural markers[4,7]. Unlike other hemangiopericytomas, SNHP has a more regular cellular arrangement, fewer mitoses, less cellular heterogeneity, lower recurrence and metastasis rates, and less manifestation of CD 34[2,4]; therefore, SNHP has been classified as a different entity from other hemangiopericytomas[4,5]. SNHP usually has low proliferative indices, but a proliferative index of $\geq$ 10% has been suggested to be associated with a more aggressive neoplasm[2]. In the present case, the SNHP cells had occasional mitoses without any necrosis (Fig. 1B), and manifested only smooth-muscle actin, vimentin, CD-34 and Ki-67, rather than S100 protein. Its proliferative index was 7%, indicating a less aggressive tumor.

SNHP should be differentiated from other sinonasal neoplasms such as esthesioneuroblastoma, adenoid cystic carcinomas, squamous cell carcinoma and nasopharyngeal angiofibroma[7]. Therefore, a biopsy is recommended initially, but biopsies should be performed carefully because of bleeding tendencies. If SNHP is confirmed, wide local excision is recommended via an endoscopic approach, lateral rhinotomy[3], or craniofacial approach, most importantly, to achieve adequate negative surgical margins. In order to prevent massive surgical hemorrhage in our case, presurgical transarterial embolization was recommended. Complete endoscopic excision was performed smoothly after transarterial embolization.

The reported metastatic rates (5~10%) of SNHP are generally lower than those (12~60%) of hemangiopericytomas in other organs. Recurrence (8~53%) has been reported to precede metastasis, but most recurrences are probably ascribed to residual neoplasm after incomplete excision[4,8]. Radiotherapy and chemotherapy can be used as both adjuvant therapy and palliation for metastatic disease, and residual or non-resectable primary neoplasms[2,7]. Overall, the prognosis is good, with 5-year- and 10-year-disease-free survival rates of 74.2% and 64.4%, respectively[2]. Because our case tumor was completely excised, the possibility of recurrence or metastasis was low and the following two years was uneventful. However, we believe that the patient should be followed regularly to

exclude the possibility of recurrence or metastasis in the future.

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Original Article

鼻及鼻竇血管外皮細胞瘤之病例報告

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鼻及鼻竇血管外皮細胞瘤是一種罕見的鼻及鼻竇間質瘤，目前認為它與一般的軟組織血管外皮細胞瘤不同。一37歲男性，因間斷流鼻血3日求診於本院。理學檢查發現左側中鼻道有一顆表面平滑的紅色腫塊，經切片後證實是鼻及鼻竇血管外皮細胞瘤。磁振造影顯示該腫瘤已傾犯後方鼻中膈及兩側後篩竇，並阻塞左側上頷竇竇口造成鼻竇炎。經動脈栓塞後，在內視鏡下以微形吸絞器切除之。爾後2年，情況穩定，不再流鼻血，磁振造影追蹤顯示兩側上頷竇炎依舊，但腫瘤並未復發，然而，本個案仍須長期追蹤以排除復發或轉移的可能性。

關鍵詞：血管外皮細胞瘤、鼻及鼻竇腫瘤、血管瘤、流鼻血、泛鼻竇炎

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