Hemangiopericytoma in the Right Buccal Area

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Hemangiopericytomas (HPCs) are rare neoplasms of vascular origin that occur in the head and neck region. These tumors arise from capillary pericytes and are difficult to distinguish from other tumors of vascular origin. Characterization is dependent on histological analysis. The rare occurrence of these tumors and their variable malignant potential has limited the attempts to characterize their clinical behavior. We report a case of HPC located in the right buccal area of a young woman. Treatment involved radical tumor excision and reconstruction using a free myocutaneous flap. At four-year post-operative follow-up, the surgical wound had healed well, and there was no evidence of recurrence. Close long-term follow-up is indicated because of the high incidence of local recurrence and metastatic spread of these tumors. **(Mid Taiwan J Med 2008;13:158-63)**

Key words

hemangiopericytoma, head and neck neoplasms, mouth neoplasms

INTRODUCTION

Hemangiopericytoma (HPC) is a benign neoplasm with malignant potential. Generally, the growth and progression of this neoplasm is slow. HPC usually presents as a painless mass. The distinction between benign and malignant forms of HPC is based on the clinical course of the disease (e.g., occurrence of metastasis and recurrence). HPCs most frequently arise in the lower and upper limbs and in the brain, although it has been reported that as many as one-third of all HPCs originate in the head and neck region [1,2].

Since there are few pathognomonic symptoms and signs of malignant HPC, the definitive diagnosis of this neoplasm is based on results of histological examination. Despite the relatively benign-appearing microscopic features of this neoplasm, distant metastases have occurred in some cases after extended follow-up periods [3]. It has been suggested that all HPCs have malignant potential, and, therefore, should be excised. In this study, we report a case of HPC located in the right buccal area of a young woman. Taking into consideration the cosmetic appearance of the patient and the malignant potential of the tumor, we selected radical resection with immediate reconstruction using a free myocutaneous flap without postoperative radiotherapy as the treatment mode.

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CASE REPORT

In March 2004, a 28-year-old woman presented to our outpatient department with a painless mass in the right cheek. The mass had been present for more than 3 years and had rapidly enlarged during her pregnancy. Her medical history was unremarkable, and there was no history of trauma. Clinical examination revealed a 7 cm $\times$6 cm soft mass with local cutaneous hypervascularization in the right cheek (Fig. 1). A 5 cm $\times$5 cm granular mass with induration in the ipsilateral buccal area was also noted (Fig. 2). A computed tomographic (CT)

scan showed diffuse heterogeneous enhancement within the tumor in the right buccal area. The tumor extended from the right inferior temporal fossa to the right submandibular region. No marked lymphadenopathy was observed. The pterygoid plate, anterior portion of the right mandibular ramus, and a part of the right maxilla had been eroded by the solid tumor (Fig. 3).

Initially, the painless tumor grew slowly. It had been incidentally diagnosed during an examination for asymmetry in the cheeks in 2001. During the entire course of tumor development, no fluctuations in the size of the tumor size were observed. Furthermore, no tumor bleeding or right cheek pain was noted. In 2002, incision biopsy of the right buccal mass was performed at the dental outpatient department. Pathologic examination of the biopsy specimens revealed the presence of squamous cell hyperplasia. However, malignancy was suspected because of a rapid increase in the tumor size during her pregnancy in 2003. In March 2004, composite resection and right supraomohyoid neck dissection were performed with immediate reconstruction using an anterior lateral thigh flap for cosmetic purposes.

Gross examination revealed the presence of a 4.5 cm × 5.0 cm × 4.7 cm yellow to tan, encapsulated, elastic mass. Microscopic examination of the mass revealed diffuse proliferation of oval- and spindle-shaped tumor cells. The cells were characterized by abundant eosinophilic cytoplasm and the absence of mitotic figures. The mass exhibited a multinodular pattern. Microscopic examination revealed gaping and ramifying vascular channels in a staghorn configuration. The channels were separated by a single layer of attenuated endothelial cells and were surrounded by tightly packed tumor cells (Fig. 4). Three dissected neck lymph nodes were found to be disease-free.

There are no specific immunohistcochemical markers for HPCs [4]. In this study, immunohistochemical tests were positive for vimentin but negative for cytokeratin, epithelial

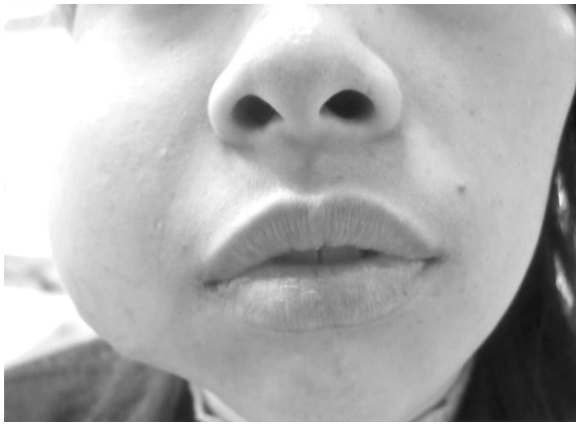

Fig. 1. A 7 cm × 6 cm soft mass with local cutaneous hypervascularization in the right cheek.

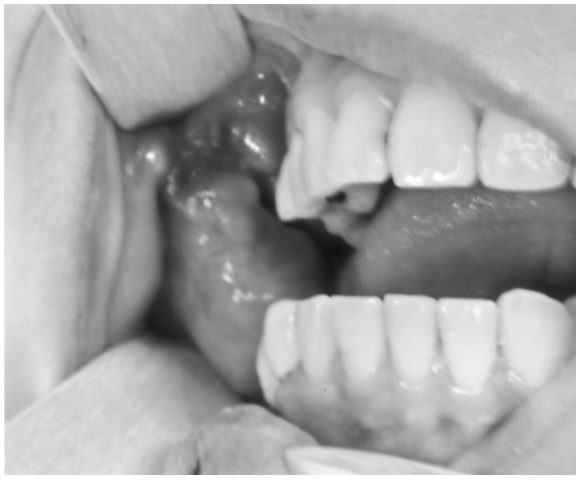

Fig. 2. A 5 cm × 5 cm granular mass with induration in the right buccal area.

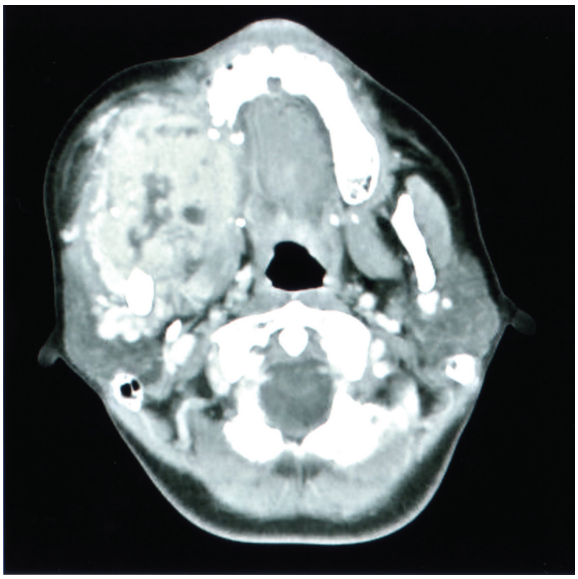

Fig. 3. Preoperative CT reveals a heterogeneous tumor in the right buccal area. The tumor extends from the right inferior temporal fossa to the right submandibular area.

membrane antigen (EMA), S-100 protein, smooth muscle actin, desmin, and CD34. The microscopic findings and results of the immunohistochemical tests indicated that the neoplasm was of mesenchymal origin.

During four years of regular postoperative follow-up in our ENT outpatient department, the operative wound healed well and there was no evidence of local tumor recurrence or distant metastasis to the lungs, skeletal system, or liver.

DISCUSSION

Pericytes were first described in 1923 by Zimmerman as smooth muscle-related cells that exhibit contractile function despite lacking myofibrils. The cells possess long processes that wrap around capillaries, thus altering the lumen caliber of these capillaries. In 1942, Stout and Murray coined the term "hemangiopericytoma" to describe tumors that consist of capillaries with their sprouts surrounded by Zimmerman pericytes. Histopathologically, these tumors are characterized by the proliferation of oval- and spindle-shaped pericytic cells around endothelial-lined vascular channels.

The architectural pattern of HPCs can also be observed in other mesenchymal neoplasms. The diagnosis of HPC is one of exclusion and relies on the detection of characteristic histological features. In the 1990s, it was reported that HPCs express XIIIa (F-XIII) and histocompatibiliy antigen HLA-DR, which were negative in most mesenchymal tumors with HPC-like patterns (including synovial sarcomas, malignant schwannomas, leiomyocoscarcoma, and liposarcoma). However, malignant fibrous histiocytoma can exhibit both F-XIII and HLA-DR [5]. Since there are no specific immunohistochemical markers for HPCs, the diagnosis of HPC is based on the exclusion of other conditions. Few immunohistochemical studies have investigated proliferation markers of HPCs, and none of them have specifically addressed the application of these markers in the diagnosis of HPCs that originate in the head and neck. For example, Middleton and colleagues [6] correctly diagnosed 7 of 9 cases of recurrent or metastatic HPCs that exhibited a proliferation rate of 15% or greater based on positive staining for MIB-1. However, none of the tumors in these cases originated in the head and neck. HPC cells occasionally express vimentin as intermediate filaments, but the intensity of vimentin staining varies considerably among tumors. Many HPCs exhibit CD34 reactivity. However, it is has been observed that HPCs are invariably negative for other endothelial markers such as the von Willebrand factor and CD31. HPCs may exhibit focal actin and desmin staining [4]. In our study, the immunohistochemical tests were positive for vimentin but negative for cytokeratin, EMA, S-100 protein, smooth muscle actin, desmin, and CD34.

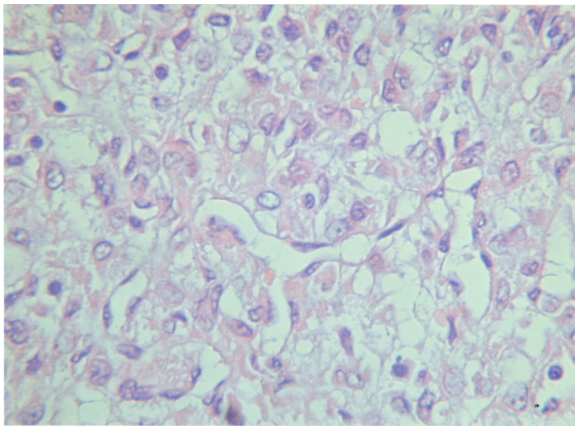

Fig. 4. Microscopic findings indicate diffuse proliferation of oval- and spindle-shaped tumor cells with abundant eosinophilic cytoplasm, a multinodular growth pattern and a staghorn configuaration.

HPC is a benign neoplasm with malignant potential. Benign and malignant forms of HPC are distinguished on the basis of the clinical course of the disease (e.g., occurrence of metastasis and recurrence). Generally, the growth and progression of this neoplasm is slow. However, the clinical features of the malignant forms are variable. These tumors are solid, elastic masses that are frequently encapsulated despite their malignant behavior. The microscopic features of ascribed prognostic value include increased cellularity, anaplasia, necrosis, hemorrhage, and prominent mitotic activity [7]. In their analysis of 106 cases, Enziger and Smith reported that 8 of 16 metastatic tumor specimens

had more than 4 mitoses per 10 high-powered field (hpf). However, in 46 of 66 HPC cases, no tumor recurrence or metastasis was observed; furthermore, no mitotic figures were observed in 30 hpfs in these cases. The remaining 8 metastatic HPCs exhibited a relatively lesser degree of mitotic activity but were hypercellular, necrotic, and hemorrhagic [7]. Similarly, McMaster et al [8] reported cases in which hypercellular, mitotically active HPCs tended to progress toward malignancy.

HPCs present as a painless mass in all age groups. It is observed predominantly in the 6th and 7th decades of life, with no sex predilection. HPCs of the head and neck can occur at any age. Their common sites of occurrence include the scalp, face, neck, oral cavity, nasal cavity, paranasal sinuses, and the orbit [9]. Although a majority of these tumors are acquired or are of the adult type, 3.3% are congenital [8]. Although trauma, prolonged steroid use, and hormonal imbalance have been shown to be associated with HPC, its etiology remains unknown. In our case, a rapid increase in tumor size was noted during the patient's pregnancy; however, there is no conclusive evidence indicating an association between HPC and pregnancy.

The incidence of regional metastasis from HPC is low, and most patients have local relapse or distant metastasis. An analysis of 45 cases of HPC of the head and neck indicated that 40% of patients relapsed locally and 10% had distant metastasis [9]. Similar results were observed in a recently conducted study in which 41% of patients relapsed locally and 33% had distant metastasis [10].

With regard to the treatment of HPCs, complete local excision is recommended for tumors that exhibit no cellular pleomorphism or necrotic or hemorrhagic areas and for those that have a uniform histological profile with no or minimal mitotic activity (< 3 mitotic figures per 10 hpf) [7]. The optimal treatment of less well-differentiated HPCs has not been clearly defined. Radical surgery may be required in some cases. Preoperative embolization or radiotherapy or both is sometimes advocated to reduce the size of the tumor and minimize the risk of operative hemorrhage. There is also evidence that supports the use of postoperative radiotherapy for these tumors [10]. Photodynamic therapy (PDT), a relatively new form of treatment, is increasingly being used for HPC. The mechanism underlying this therapy is as follows: photochemical reactions, which are initiated by the light activation of photosensitizing drugs, result in tumor cell death. Tissue damage induced by PDT is healed mainly by tissue regeneration with no scarring. Moreover, PDT does not severely impact important anatomical structures or functions and has a good cosmetic outcome [4]. However, owing to the limited penetrating depth of light, PDT is usually used for superficial or mucosal lesions. Because of the cost and anatomical limitations, PDT therapy for head and neck lesions is not generally considered a suitable mode of treatment in Taiwan. In our case, the patient did not present with poor prognostic factors such as large tumor size (diameter > 6.5 cm), mitotic activity, necorsis, or hemorrhage. Taking into consideration the cosmetic appearance of the patient and the malignant potential of the tumor, we selected radical resection with immediate reconstruction using a free myocutaneous flap without postoperative radiotherapy as the treatment mode.

Backwinkel and Diddams [11] reported that the overall recurrence rate of HPC was 52.2%. The recurrence rate of HPCs in the central nervous system, the head and neck, and in the uterus were 80%, 57.1%, and 41%, respectively. This high recurrence rate can be explained by the fact that although these tumors are encapsulated, they variably exhibit an invasive growth pattern [12].

The interval of HPC recurrence is often long or delayed. Enzinger reported that in 7 of 16 patients, HPC recurred after a disease-free interval of 3 years [7]. In our patient, there was no evidence of local recurrence or distant metastasis during the 4 years of regular postoperative follow-up conducted in our outpatient clinic. Because of the high incidence of the postoperative recurrence of HPC, long-term follow-up is important.

In summary, the relatively low incidence of HPCs and their unpredictable behavior limit our ability to establish an optimal treatment strategy. The preferred treatment is wide surgical excision [6,10]. Patients with large lesions (> 6.5 cm in diameter), necrosis, positive surgical margin, and high-grade lesions (> 3 mitotic figures per 10 hpf) may benefit from adjuvant external-beam radiation therapy [7].

In our experience, radical surgical resection alone for HPCs in the head and neck can provide good local control. However, long-term follow-up is important because of the high incidence of the postoperative recurrence of HPC. Although distant metastasis in HPCs is quite rare, it should be considered during the treatment of this neoplasm.

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右頰部血管外皮細胞瘤

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血管外皮細胞瘤爲頭頸部罕見腫瘤之一，此腫瘤多爲生長緩慢、界線分明之血管性腫瘤，治療以外科手術切除爲主，但其復發率高。在此提出之病例爲一28歲女性，主述爲期兩年右側頰部無痛性逐漸腫大之腫塊，經診視發現右側頰部有些許的肉芽組織併巨大深部腫塊，右頰部局部切片檢查並無惡性變化，電腦斷層檢查顯示一不均質之大腫塊位於右側頰部之顳窩與下頷骨之間。患者於93年3月接受右頰部腫瘤之廣泛性切除與前大腿游離皮辦顯微重建手術，病理報告證實爲血管外皮細胞瘤，術後傷口復原狀況良好，術後4年追蹤電腦斷層檢查顯示並無局部復發之徵象。

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關鍵詞

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