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A Case of Malignant Hemangiopericytoma in Face

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안면부에 발생한 악성 혈관외피세포종 1례

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= 국 문 초 록 =

혈관외피세포종(Malignant hemangiopericytoma)은 난원형의 혈관외피세포(pericyte)들로 구성된 매우 드문 혈관종의 일종으로 대개 사지 및 후복막에 발생되며, 안면부와 경부에도 약 25% 가량의 발생율이 보고되어 있다. 성별 발생빈도의 차이는 없고 20~50대에서 호발하며, 종양은 대부분 서서히 성장하고 경계가 명확한 무통성의 고형 종괴로 나타난다. 악성과 양성의 구별은 조직학적 소견과 주변 조직으로의 침습, 원격전이 여부 등을 고려하여 판단하지만, 양성으로 진단된 경우에도 수년 내에 재발 및 원격전이가 나타나는 경우가 다수 보고되므로 악성과 양성의 감별이 매우 어렵고, 양성인 경우에도 악성 잠재성이 존재하는 것으로 알려졌다. 치료는 수술적 완전절제가 유일한 방법이나 절제가 불완전한 경우는 수술 후 외부 방사선 조사를 한다. 항암제 투여는 효과면에서 논란이 있지만, 원격전이나 수술과 방사선 치료가 실패한 경우 시행할 수 있다.

저자들은 최근 안면부에 발생한 악성 혈관외피세포종 환자 1예를 치험하였기에 보고하는 바이다.

중심 단어 : 혈관외피종 · 안면부.

Introduction

Hemangiopericytoma is an uncommon vascular tumor of unknown etiology[1]. About 25% of these tumors occur in the head and neck[2]. Clinically, the tumor may occur at any age, with the highest incidence between the third and sixth decades and without any sex predilection[3-5]. These lesions are characteristically slow-growing and slow to metastasize[6-9]. However, regarding the late recurrence and metastasis, many authors consider them as a malignancy[10][11]. The treatment of choice is radical removal of the tumor, but it may often not be possible[12]. Radiation therapy and chemotherapy are justified only as an adjuvant measure[10][12]. We present the case of 49-year-old Korean man with hemangiopericytoma who was treated by radical removal of the tumor and postoperative radiation therapy.

Case History

A 49 year-old-man was initially seen with a 2-month history of a rapidly growing mass in the right

cheek. The patient had been diagnosed mycosis fungoides at age 29 and had undergone radiation therapy. Four years later, he was also diagnosed malignant lymphoma and treated with chemotherapy. On physical examination, there was a firm, well marginated, round and non-tender mass on his right cheek. The mass was fixed to the surrounding tissue(Fig. 1). All of the branches of right facial nerve were weakened, especially the buccal and marginal mandibular branches. Computed tomographic scan of the face showed a well-circumscribed, homogeneously enhancing mass in the right masticator space with infiltration into the masseter muscle. There were some enlarged lymph nodes on the submandibular area and jugulo-digastric area, but all of them seemed to be reactive hyperplasia(Fig. 2). The incisional biopsy of the mass showed non-hematogenous malignant tumor of uncertain epitheloid feature. In December 1997, the patient underwent wide excision of the cheek mass, partial resection of the masseter muscle, and superficial parotidectomy. As well, some minor branches of buccal branch were sacrificed during the operative procedure. The specimen consisted of a well-circumscribed solid mass with a smooth, glistening external surface. Multiple cut section revealed a myxoid and fleshy appearance with focal hemorrhagic necrosis. Histological evaluation revealed a non-hematologic sarcomatous feature enriched with vasculature lined

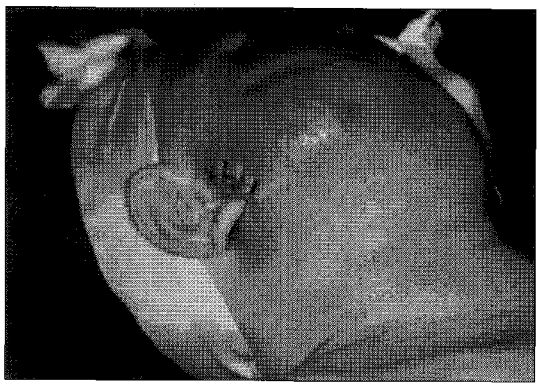

Fig. 1. Photography of face after the incisional biopsy.

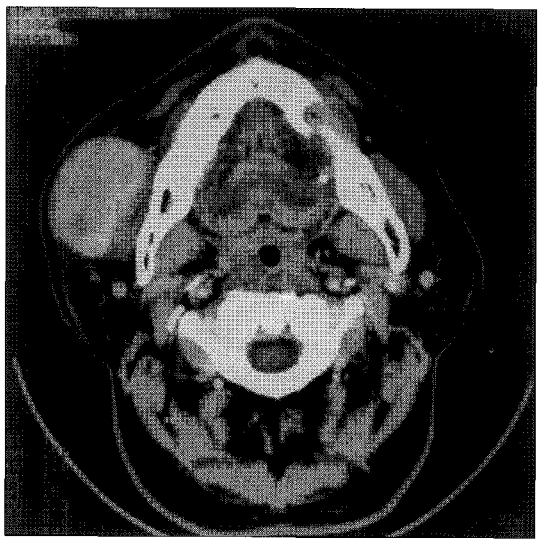

Fig. 2. CT scan of the lesion : well-circumscribed homogeneously enhancing mass in the right masticator space with infiltration into the masseter muscle. There were some enlarged lymph nodes on the submandibular area and jugulo-digastric area, but all of them seemed to be reactive hyperplasia.

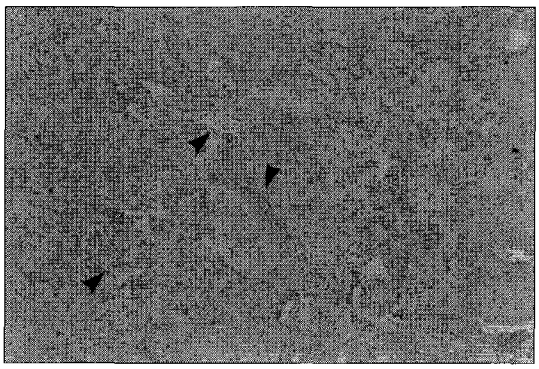

Fig. 3. Histologic findings(H-E stain 100) : non-hematologic sarcomatous feature enriched with vasculature lined by flattened endothelium. The vasculature showed a distinct stag-horn pattern.

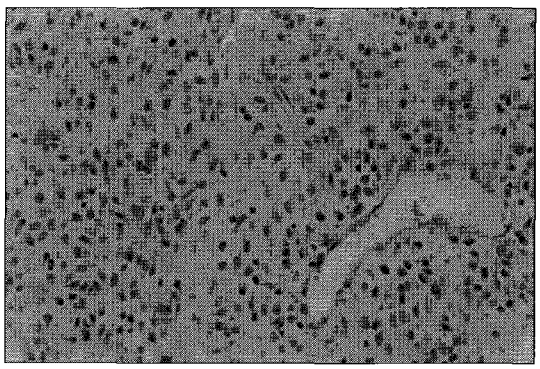

Fig. 4. Histologic findings(H-E stain 200) : dense cellular component of matrix which is composed of malignant cells showing occasional mitotic figures.

by flattened endothelium. The elongated tumor cells had round or ovoid nuclei and mitotic figures were frequently seen(Fig. 3, 4). The immunohistochemical stain demonstrated a strong positive reaction with vimentin and partially positive to actin and desmin, but no positive reaction of tumor cell with CD34 and CD31. After the operation, local radiation therapy(6400cGy) was given. The patient has remained free of disease following surgery.

Discussion

Hemangiopericytomas are uncommon vascular tumors consisting of diffuse elongated cells arising from pericytes enclosed in reticular fibers, first described in 1942[1]. These tumors occur most often in extremities and retroperitoneum, although 13% to 25% are found in the head and neck[2]. Hemangiopericytomas have varied courses, but the usual pattern is of indolent growth[12]. Owing to the slow growth, these tumors have sometimes been misdiagnosed as lipomas. The recurrence rate of these diseases has been reported as more than 50%, and the metastasis rate ranging from 12% to more than 50%[6-9]. Malignant hemangiopericytomas can be usually diagnosed by application of the criteria of Enzinger & Smith[13]. The occurrence of delayed metastasis occasionally can be seen even in patients with histologically and cytologically benign looking hemangiopericytoma makes the existence of benign hemangiopericytomas questionable[10][14][15]. It appears appropriate to consider all hemangiopericytomas as potentially malignant. Some authors advocated that an observation period is at least 10 years required before a fairly reliable statement can be made about the behavior of the tumor[10].

The radical removal of the tumor has been recommended as the treatment of choice, but, due to the anatomical configuration, complete excision may not be always feasible. Preoperatvie angiography with simultaneous embolisation by microparticles or platinum minispirals may be helpful in reducing the size of the tumor before surgery as well as reducing intraoperative hemorrhage and the chance of intraoperative scattering of tumor cells[10]. Postoperative radiotherapy is recommended at least in the cases of incomplete surgery, although these tumors are thought to be fairly resistant to radiotherapy[6]. If surgical and radiotherapeutic means are exhausted, chemotherapy may also be considered[16][17] even though some authors believe that the chemotherpy is not effective in patients with hemangiopericytoma[5][18].

We suggest that it would be mandatory to follow-up patients with hemangiopericytoma for life time to identify any recurrence or late metastasis.

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