Hemangiopericytoma of the parotid gland: A case report

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Hemangiopericytoma is an uncommon tumor of vascular origin arising from the pericytes of Zimmerman. It represents only 1% of all vasoformative tumors. Up to one fourth of all cases of hemangiopericytoma are reported from head and neck region. An origin in the parotid gland is extremely rare, with only 30 cases reported in the world literature. Because of the rarity of the condition and its unusual site of origin, a case of hemangiopericytoma of the parotid gland is being reported in a 55-year-old woman. [Turk J Cancer 2000;30(2):89-93]

Key words: Hemangiopericytoma, parotid gland

The term hemangiopericytoma was first coined by Stout and Murray in 1942. It is a rare mesenchymal tumor which originates from the pericytes of Zimmerman surrounding the capillaries and post capillary venules. It constitutes only 1% of all vasoformative tumors of the body (1). Trauma, steroid therapy and prolonged chemical exposure have been linked with its etiology. Though, it has been reported to occur at any age from birth to 92 years, it is primarily a tumor of adults and rarely found in children. It is equally found in either sex. Although it can be found throughout the human anatomy, the lower extremities and the retroperitoneum/pelvis are the most common sites. The head and neck region is the third commonest site for hemangiopericytoma (2).

Case Report

In January 1995, a 55-year-old woman presented with a 5-year history of painless mass on the right side of face which had been excised twice in another hospital. The last removal of the swelling, performed about 18 months before attending the Cancer Institute was without any information about the histology. The present swelling was slowly increasing since last surgery and has rapidly
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increased in size for the last one month. Physical examination showed a 6x5 cm size firm to hard, non-tender mass in the right pre-auricular region. The skin overlying the swelling had a 3 cm long scar. The skin was tense but local temperature was normal. No evidence of facial weakness or cervical lymphadenopathy was present. Systemic clinical examination and routine laboratory investigations, including hemogram, urine examination and chest X-ray were normal. Fine needle aspiration cytology of the lesion was inconclusive. CT scan of the head showed a 52x45 mm sized soft tissue mass having irregular margins in the right parotid gland region and showing homogenous enhancement after contrast injection (Figure 1). No calcification within the mass or erosion of underlying bone was noted. CT scan was suggestive of a malignant mass in the right parotid gland.

Fig 1. CT scan of the head showing densely enhancing right parotid mass.

The patient underwent a right superficial parotidectomy with total removal of the mass. The underlying facial nerve was preserved. Gross examination of resected specimen revealed salivary gland with tumor weighing 35 g and measuring 6.0x5.0x4.5 cm in size. On sectioning, a tan, smooth, firm, circumscribed tumor measuring 4x3 cm was found in the salivary gland. Microscopically, the tumor had almost replaced the superficial lobe of the parotid gland, being rich in capillaries and small vessels which were lined by a single layer of endothelial cells around that were tightly packed but diffusely arranged cells (Figure 2). The cells were round to oval to spindle shaped with mild pleomorphism, vesicular nuclei and inconspicuous nucleoli. Cytoplasm was moderate in amount, with indistinct cell borders. Mitosis was observed to be 0-1 per 10 hpf. No evidence of hemorrhage or necrosis was present. The histology was suggestive of hemangiopericytoma of the parotid gland. A silver reticulin stain showed reticulin fibres that surrounded the tumor cells outside vascular spaces. Immunohistochemistry showed that the tumor cells were positive for
Vimentin but negative for Cytokeratin (CK), Epithelial membrane antigen (EMA), Smooth muscle actin, S-100 protein, and factor VIII related antigen. A final diagnosis of hemangiopericytoma of the parotid gland was made.

Fig 2. (A) low power view of the hemangiopericytoma showing ramifying vascular channels lined by single layer of flattened endothelial cells being clearly separated from tumor cells by basement membrane (H&E stain x10). (B) Dilated sinusoidal vascular channels surrounded by tumor cells with indistinct cellular outlines (H&E stain x40). (C) Silver reticulin preparation reveals dense reticulin meshwork surrounding vessels and tumor cells (x40).

Post-operatively, the patient received radiotherapy. A total dose of 5000 cGy/25 fr./5 week was delivered by 8 Mev electron beam on linear accelerator. Currently, the patient remains without evidence of local recurrence or distant metastasis 60 months following treatment.

**Discussion**

According to Batsakis and Rice (3), hemangiopericytomas located in the head and neck region account for 15 to 25% of all cases. The meninges, the nasal passages and paranasal sinuses, and the orbit are the three major sites for head and neck hemangiopericytoma (2). A location in the parotid gland is exceptionally rare. The first case of hemangiopericytoma of the parotid gland was reported by Stout in 1956 (4). A review by Pagliaro et al. (5) showed that till 1988, only 16 cases of hemangiopericytomas of the parotid gland had been reported. A search of the more recent literature revealed further 14 cases reported as single case reports or small series, making a total of 30 cases (6-
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12). The present case is the first being reported from India. Because of rarity of this tumor, it practically never enters into the clinical differential diagnosis of the parotid tumors.

Hemangiopericytoma is characteristically a very slow growing lesion and majority of the tumors are painless, thus delay in presentation and diagnosis is a common feature. It is often large in size at the time of diagnosis (13). A very slow growing painless mass over the years in the preauricular region is the commonest finding of hemangiopericytoma of the parotid. Because of the rich vascularity, telangiectasias and raised temperature of the overlying skin may occur. Other signs such as pulsation and an audible bruit may also be noted (10).

Histologically, both benign and malignant variant of this tumor have been reported. The histopathological criteria for differentiating the benign and malignant hemangiopericytoma have been mentioned by McMaster in 1975 and later by Enzinger and Smith in 1976 (13,14). Nevertheless, the biological behaviour of this tumor is quite unpredictable, and even histologically benign tumors can behave aggressively and have capacity to metastasize. Thus, conventional histological criteria for benign and malignant lesions do not always apply to this tumor that all cases of hemangiopericytoma should be considered as potentially malignant (7).

Further, many soft tissue sarcomas can mimic the histologic picture of hemangiopericytoma. Thus, diagnosis in difficult situation can be made by excluding other lesions by various immunohistochemical markers for those tumors as there are no antibodies specific to hemangiopericytoma (15).

Local recurrence and distant metastases are common features of hemangiopericytoma. Lung and bone are the most common sites for distant metastases (13). Distant metastases are reported in 11 to 56% cases (16,17). Lymph node metastases are rare.

Though wide surgical resection is the treatment of choice for these tumors, local recurrence rate following surgery alone is high (20 to 50%). Despite the fact that hemangiopericytoma is a radiosensitive tumor, radiotherapy has been used mainly for advanced and inoperable, locally recurrent/metastatic tumors (18,19). Adjuvant post-operative radiotherapy reduces the incidence of local recurrence (20). The role of chemotherapy is inconclusive. Among 30 cases of hemangiopericytoma of the parotid gland mentioned in the literature, a sufficient follow up is lacking in 13 cases, and 9 patients died after an interval varying from a minimum of 16 months to a maximum of 11 years and 5 months. Many had local and distant recurrence. Only 8 patients were disease free at 1 to 16 years of follow up. The lesion in our patient had recurred twice at local site, possibly due to incomplete excision. But, there has been no recurrence or distant metastasis at 60 months of follow up after definitive surgery and post-operative radiotherapy.

Hemangiopericytoma is known to recur locally after years or even decades following initial treatment. Latency period up to 33 years has been reported. The patient should be followed up for the life time and a minimum period of at least 10 years after initial treatment has been suggested before labelling the patient as "cured" (7,20).
References