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CASE REPORT

CT imaging of malignant metastatic hemangiopericytoma of the parotid gland with histopathological correlation

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Abstract

We report an extremely rare case of malignant hemangiopericytoma (HPC) of the parotid gland and its metastatic spread to lung, liver, and skeletal muscle. Computed tomography (CT) imaging, histopathological and immunohistochemical methods were employed to study the features of malignant HPC and its metastases. CT imaging was helpful to determine the exact location, involvement of adjacent structures and vascularity, as well as evaluating pulmonary, hepatic, peritoneal, and muscular metastases. Immunohistochemical and histopatholgical features of the primary tumor as well as the metastases were consistent with the diagnosis of malignant HPC.

Keywords: *Malignant hemangiopericytoma; parotid gland; CT imaging; metastases; histopathology.*

A 56-year-old Chinese woman presented with a progressively enlarging painless swelling in the left preauricular region. Her medical history was unremarkable. Physical examination showed a firm, immobile, non-tender palpable mass. No enlarged cervical nodes were felt. There was no neurological involvement. A computed tomography (CT) scan revealed a well-defined soft tissue mass measuring about 5 cm in diameter arising from the superficial lobe of the left parotid gland (Fig. 1). The mass was hyperdense relative to the normal parotid parenchyma on unenhanced scans and showed intense homogeneous enhancement after intravenous iodinated contrast administration. It extended superiorly over the pre-tragal area and over the left zygomatic arch. Medially, it abutted the ramus of the mandible. However, there was no associated bone destruction. There was no invasion into the deep lobe of the parotid gland, infiltration of the masticator space or the overlying skin. No enlarged cervical lymph nodes were detected. Fine needle aspiration cytology revealed pleomorphic spindle cells forming vascular spaces, raising the possibility of hemangiopericytoma (HPC). Left parotidectomy was performed. At surgery the tumor was seen to arise from the superficial lobe of the parotid and was adherent to the zygomatic arch.

The temporal and zygomatic branches of the facial nerve were stretched but were not infiltrated by the tumor. Other branches of the facial nerve were intact. Gross examination of the specimen showed a well-circumscribed pale yellow nodule measuring $5.5 \times 2.8 \times 3$ cm within the superficial lobe. Histopathological studies revealed markedly pleomorphic spindle cells forming vascular spaces, some of them slit-like and gaping, and with an antler-like appearance (Fig. 2a; high power). A rare focus of necrosis was also seen. Abnormal mitosis was noticed, the mitotic content being approximately 13 per 10 high-power fields (Fig. 2b). Immunoperoxidase staining showed the absence of S100 and glial fibrillary acidic protein (GFAP) which are usually positive in myoepithelial tumors. However, vimentin, smooth muscle actin and CD34 were positive. These features were consistent with a malignant HPC.

CT scans of the chest and abdomen were also performed at this time to rule out the presence of any distant metastases in view of the histopathological diagnosis. The patient was kept on close follow-up since local recurrence as well as distant metastases are known to occur. After surgery, the patient was disease-free for nearly 5 years. She subsequently developed pulmonary and hepatic
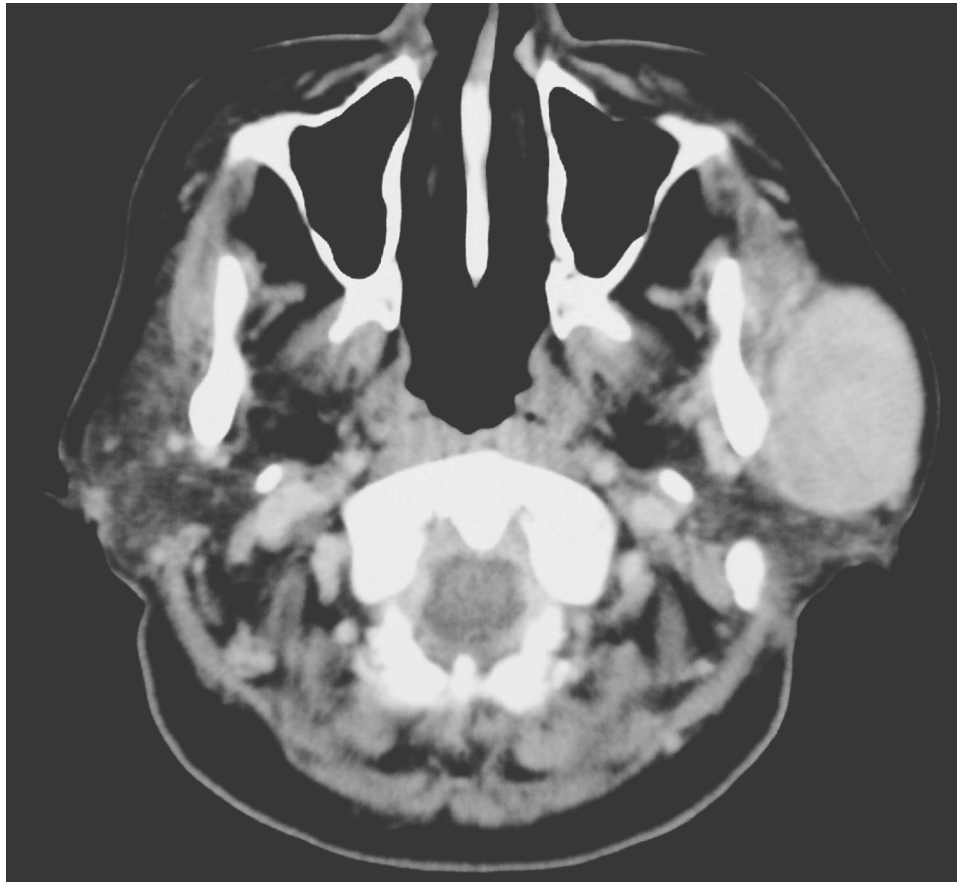
Figure 1 **CT scan revealed a well-defined soft tissue mass measuring about 5 cm in diameter arising from the superficial lobe of the left parotid gland.**
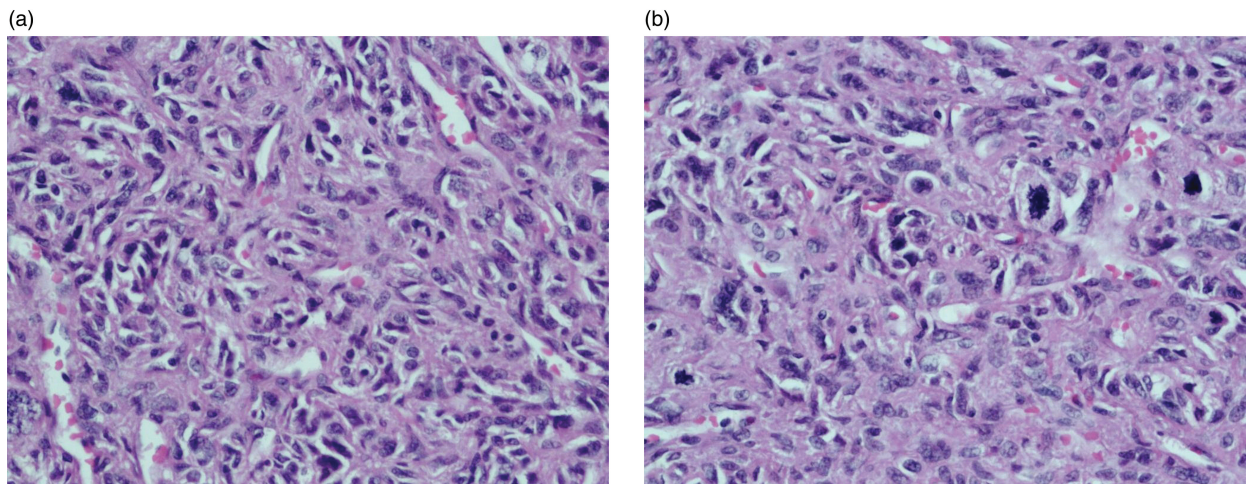
(a)
(b)

Figure 2 **(a) Markedly pleomorphic spindle cells forming vascular spaces, some of them slit-like and gaping, and with an antler-like appearance. A rare focus of necrosis was also seen. (b) Abnormal mitosis was noticed, the mitotic content being approximately 13 per 10 high-power fields.**

metastases. CT demonstrated a pulmonary mass in the right lower lobe (Fig. 3a) and hepatic metastasis (Fig. 3b). The patient underwent surgical excision of the lung and liver metastases. Histopathologic studies revealed pleomorphic ovoid to spindle shaped tumor cells with vesicular nuclei, inconspicuous nucleoli and poorly defined cytoplasm. Numerous mitoses, including abnormal forms were present. The tumor cells were

Figure 3   **(a) CT demonstrated a pulmonary mass in the right lower lobe and (b) hepatic metastasis. (c) CT scan of the pelvis showed enhancing masses in the gluteus maximus muscle confirming metastatic spread.**

positive for vimentin and negative for CEA, smooth muscle antigen and hepatocyte antigen. Factor VIII and CD34 were also positive thus highlighting the intimate relationship of blood vessels with the tumor. These features are in keeping with metastatic tumor. She further developed soft tissue swellings in the buttocks about 6 months after resection of the pulmonary and hepatic metastases. CT scan of the pelvis showed large masses and enhancing nodules in the gluteus maximus muscle and in the peritoneal cavity confirming metastatic spread (Fig. 3c).Further imaging of the chest and abdomen revealed multiple large masses involving the bowel mesentery and the peritoneal cavity as well as numerous metastases in lung and liver.

HPCs are uncommon vascular tumors arising from perivascular cells known as 'pericytes' located outside the basement membrane of the capillary wall. The term hemangiopericytoma was first introduced by Stout and Murray to describe such tumors[1]. Pericytes possess smooth muscle cell characteristics and are supposed to play a role in the regulation of blood flow through their contractile ability. On the basis of electron microscopic studies, it has been postulated that these cells are derived from primitive mesenchymal precursors and represent a transitional form between mesenchymal and smooth muscle cells[2]. HPCs constitute 1% of all vascular tumors and can be found in all age groups ranging from 10 to 65 years. The most common sites for HPCs include the lower extremities, pelvis, retroperitoneum, trunk, head and neck[3,4]. In the head and neck region, the most common locations are the nasal cavity, paranasal sinuses and orbits; HPC is extremely rare in the parotid glands and temporal bone[5–8].

Clinically HPCs present as painless slow growing tumors and almost never cause neurological symptoms or adenopathy. Diagnostic work up includes CT and/or magnetic resonance (MR) imaging. In our case CT imaging showed a well-defined soft-tissue mass located in the superficial lobe of the left parotid gland which was relatively hyperdense with respect to the normal parotid gland. On contrast studies, the mass displayed intense homogeneous enhancement. No enlarged cervical nodes were identified. On MR imaging the tumor had signal characteristics consistent with any soft tissue tumor with high signal intensity on T2-weighted images and intermediate signal intensity on T1-weighted images.[8] Homogeneous enhancement is seen on post gadolinium T1-weighted scans. Differential diagnosis based on imaging of an enhancing lesion arising from the parotid gland would include pleomorphic adenomas and mucoepidermoid carcinomas. Large pleomorphic adenomas may have lobulated or poorly defined margins; high grade mucoepidermoid carcinomas are poorly defined with heterogeneous internal architecture and may have associated cervical adenopathy.[9] Imaging shows the tumor extent and invasion of adjacent structures — information that is critical for surgical planning.

Histopathological and immunohistochemical findings form the basis for the diagnosis of HPC[3,7]. Battifora described three types of histological pattern for HPC.[9] In the first type, plump, ovoid, or round tumor cells intimately surround the capillaries. The capillary lumen is open, the reticulin sheath delicate, and collagen scant. The second category describes spindle-shaped cells around open, round capillaries with thick collagen not only separating the endothelial cells from the tumor cells, but also the individual tumor cells. The last category exhibits spindle-shaped cells uniformly distributed along very narrow and often inconspicuous capillary lumina associated with poorly visualized basement membranes and pleomorphism of the endothelial cells. The histological pattern in our case resembles the last category in all respects. However, the last type of histological pattern can be mistaken for the diagnosis of vascular soft tissue sarcomas as these tumors may show areas of 'hemangiopericytoma-like' vascularity[10].

McMaster *et al.* used the histological pattern to differentiate benign and malignant tumors and found that 32 of 60 cases were malignant (53%), with a 5-year survival rate of 25%[11]. Enzinger and Smith showed that a higher rate of mitotic activity, increased cellularity, and the appearance of undifferentiated cells, necrotic and hemorrhagic regions in the tumor tissue are common signs of malignancy and thus have increased chances for recurrence or development of metastases[4]. Better survival rates (10 years) of patients were reported for tumors with fewer mitoses (<4 mitotic figures/10 hpf), the absence of necrosis, and size less than 6.5 cm[1,4]. The lesion with higher mitoses (>4 mitotic figures/10 hpf) is invariably malignant and has greater tendency to metastasize compared to the tumor with fewer or no mitoses[5]. The higher mitotic figures (13/10 hpf) and the presence of necrosis in our patient suggests greater malignant potential. Malignancy was also reported for tumor size below 6.5 cm (4–6 cm) in some anatomic locations and is consistent with the size (5 cm) seen in our case[3,5]. Immunohistochemical features and histopathological features such as pleomorphism, hypercellularity, higher mitotic rate, and hypervascularity are in accordance with those reported for malignant HPC[4–8]. S100 and GFAP were used to exclude other mesenchymal neoplasms[4]. Our patient developed metastases in the lungs and liver 5 years after the surgery. Distant hematogenic spread preceded local recurrence in two-thirds of patients with metastases and the time interval for metastasis is about 5 years in 30% of the cases[3]. According to Noltenius, a malignant HPC was clinically characterized by the presence of hematogenic metastases, typically to the lungs, liver and skeletal system[12]. CT in our case confirmed this type of metastatic spread to the lungs and liver. Thoracotomy was performed in our patient for resection of lung metastasis and segmental resection was carried out for removal of hepatic metastasis. Soft tissue swellings were noticed in the buttocks 6 months

from the time of resection of lung and liver metastases. CT of the pelvis revealed metastases in the glutei muscles. Repeat CT scans of the thorax and abdomen also showed numerous metastases in lung, liver and abdomen.

This report describes the long-term follow-up of a patient with a rare malignant HPC arising from the parotid gland which later metastasized to lung, liver, mesentery and skeletal muscle. CT was helpful in determining the precise location, involvement of adjacent structures, and the vascularity of the primary tumor in the parotid gland as well as pulmonary, hepatic, peritoneal and intramuscular metastases. Histopathological and immunohistochemical features are consistent with a diagnosis of malignant HPC. This study also emphasizes the need for long-term follow-up after surgery in such tumors.

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