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CASE REPORT

Hemangiopericytoma of the parotid gland: a case report

Parotis bezinde saptanan hemanjiyoperisitoma: Olgu sunumu

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Hemangiopericytomas are uncommon neoplasms of vascular origin that mostly arise from extremities, retroperitoneum, and pelvic fossa. They are rarely found in the parotid gland. A 35-year-old female patient presented with a swelling in the right preauricular region. Fine-needle aspiration biopsy performed twice yielded an extraordinary amount of blood. A cytological diagnosis could not be made. Computed tomography showed a homogenous, well-shaped mass in the parotid gland. Superficial parotidectomy was performed. Histopathological diagnosis was made as hemangiopericytoma. No evidence for local recurrence or distant metastasis was seen within a follow-up of 36 months.

Key Words: Hemangiopericytoma/pathology/surgery; parotid gland.

Hemanjiyoperisitoma daha çok ekstremiteler, retroperiton ve pelvik fossa, nadiren de parotis bezinde gelişen vasküler bir tümördür. Otuz beş yaşındaki kadın hasta sağ preauriküler bölgede şişlikle kliniğimize başvurdu. İki kez uygulanan ince iğne aspirasyon biyopsisinde olağan dışı miktarda kan geldiği görüldü. Sitolojik tanı konulamadı. Bilgisayarlı tomografide parotis bezinde yerleşmiş, iyi sınırlı kitle saptandı. Yüzeyel parotidektomi ile tümör tamamen çıkarıldı. Histopatolojik incelemede hemanjiyoperisitoma tanısı kondu. Hastanın 36 ay içinde yapılan kontrollerinde lokal nüks veya uzak metastaza rastlanmadı.

Anahtar Sözcükler: Hemanjiyoperisitoma/patoloji/cerrahi; parotis bezi.

Hemangiopericytomas are rare mesenchymal vascular tumors that originate from the pericytes around the capillaries. These are mostly seen in extremities, pelvic fossa, retroperitoneum, head and neck region, although they can develop anywhere around capillaries.[1] Hemangiopericytomas constitute 1% of vascular masses and 13-25% of them are localized in the head and neck region.[2] In the head and neck region, they are mostly seen in the nasal cavity and paranasal sinuses but rarely arise from the parotid gland.[3] They are mostly seen in the fifth and sixth decades. There is no difference between sexes in terms of prevalence. Hemangiopericytomas may show different growth patterns, clinical behaviours and malignancy potentials. Radiological imaging studies are not sufficient for the diagnosis. In this article we report a localized mass in the parotid gland that was treated with superficial parotidectomy. Histopathological diagnosis was hemangiopericytoma.

CASE REPORT

A 35-year-old female patient presented with a swelling in the right preauricular region of a four-year history. There was no enlargement at all for the

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past years. Fine-needle aspiration biopsy performed twice yielded an extraordinary amount of blood. A cytological diagnosis could not be made. Ultrasonography revealed a hypoechoic vascular mass of 24x24 mm in the superficial lobe of the parotid gland. Computed tomography (CT) showed a homogeneous, well-shaped mass in the parotid gland measuring 3x2.5x3 cm. It was close to the external ear canal (Fig. 1). The hemogram, biochemical tests, and serology of the patient were normal.

Superficial parotidectomy was performed. The tissues were very fragile and blood loss was more than expected. The resected material was about 3x2x1 cm in size, near-round in shape, elastic-hard in consistency. Its cut surface had a fibrous appearance. Tumor tissue was composed of partially spindle cells that showed sinusoidal clefts and formed blunt fusiform bundles (Fig. 2) These cells were oval or round in nature, some of them had fusiform or cuboidal cytoplasm. The number of mitoses per 10 high-power fields was 1. Necrosis was not found. The differential diagnosis included vascular origin tumors (hemangioma, hemangioendothelioma, hemangiopericytoma) because of similar morphological findings, glomus tumor and fibrous histiocytoma due to fusiform proliferation, and myoepithelioma due to localization. Pancreatin, CD31, CD34, smooth muscle actin were used for immunohistochemical investigation, and Gomori's reticulin stain was used for conventional histochemistry. In reticulin staining, black reticulin fibers were found typically covering tumor cells and vascular structures. While the tumor cells were stained negatively with pancreatin, smooth muscle actin and CD31, a focally weak positivity was found for CD34 (Fig. 3, 4). Considering morphological findings, immunohistochemical staining properties and reticulin patterns, the diagnosis was made as hemangiopericytoma.

During 36 months of follow-up, there was no evidence for local recurrence or distant metastasis.

DISCUSSION

Hemangiopericytoma was first defined by Stout and Murray in 1942. Post-trauma, steroid use, and genetic factors have been implicated as etiologic causes, but none has been ascertained. Histologically, the tumor is characterized by capillaries that are surrounded by small oval cells (Zimmermann pericytes). Diagnosis of hemangiopericytoma is difficult because of similar distribution of many soft tissue tumors (monophasic fibrous synovial sarcoma, mesenchymal chondrosarcoma, infantile fibrosarcoma, malignant fibrous histiocytoma, malignant schwannoma, leiomyosarcoma, malignant mesothelioma, solitary fibrous tumor).[4]

Despite the notification of vimentin positivity in hemangiopericytoma, it is not specific and shows differences from one tumor to another. Contrary to other vascular tumors, hemangiopericytoma cells do not show positive staining for factor VIII-related antigen,

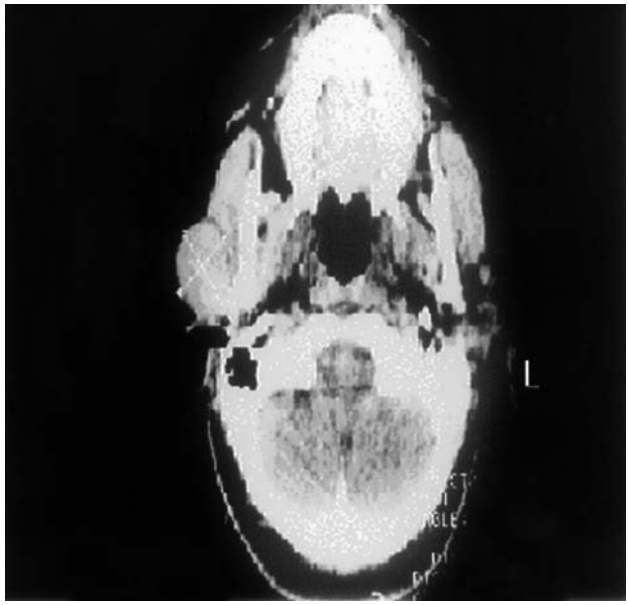

Fig. 1 - *Parotid mass in the axial computed tomography scan.*

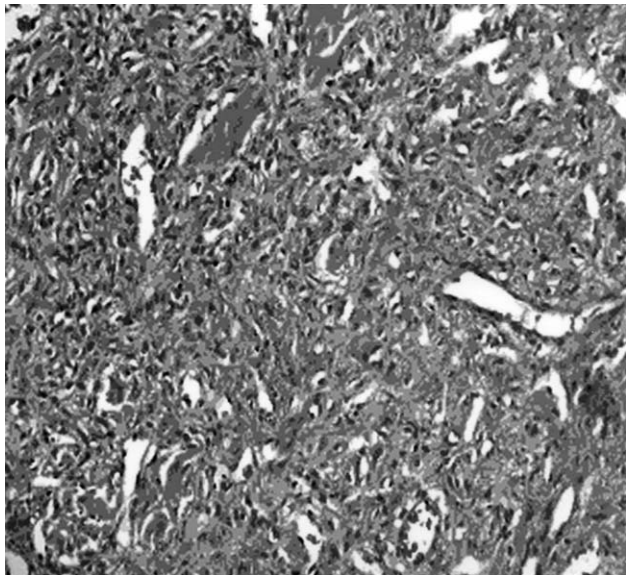

Fig. 2 - *Partially spindle cells showing sinusoidal clefts and forming blunt fusiform bundles (H-E x 40).*

CD31, Ulex europaeus I lectin, or smooth muscle actin.[5] But they show CD34 positivity similar to other vascular tumors. It is hard to differentiate between benign and malignant forms. Although hemorrhage, necrosis, and increased cellularity suggest malignancy,[2] the number of mitoses is considered to be one of the most important parameters of clinical behavior.[5] Therefore, the presence of 2-3 or less mitoses per 10 high-power field is a positive prognostic factor, and the presence of four or more mitoses is associated with recurrence and metastasis due to faster growth capacity. However, cellular anaplasia and high mitotic index may not always be reliable, because metastatic cases despite low mitoses were reported.[1]

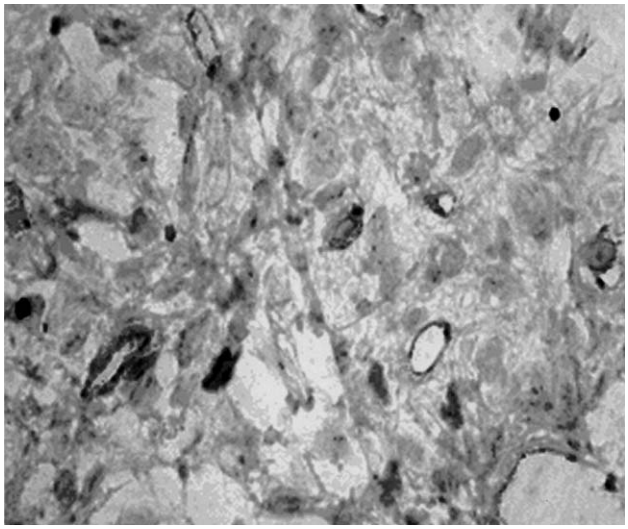

Fig. 3 - *Tumor cells showing negative staining for CD31 (x200).*

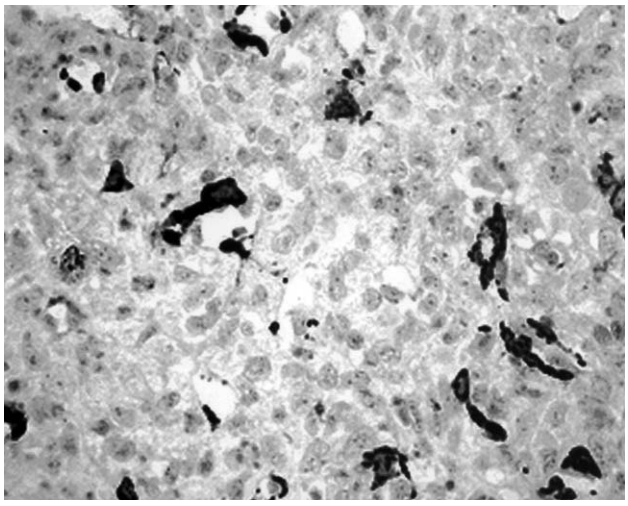

Fig. 4 - *Negative staining for smooth muscle actin in tumor cells but positive staining in the vessels around the tumor cells (x200).*

Although it is not certain that tumor size affects malignant behaviour, tumors greater than 5 cm were reported to be associated with malignant behaviour.[1] These may show an adverse histological behavior and 50% are malignant in nature. While tumors in elderly patients may show a malignant course, congenital, infantile forms, and superficial tumors are benign in nature. Recurrence is a manifestation of malignancy. Recurrences even after 28 and 33 years were reported.[6] Regional lymph node involvement is rare and is most commonly associated with metastasis to the lungs, bone, and liver. Since metastasis may occur long after the resection of primary tumor, lifelong follow-up is necessary. Even the benign tumors may metastasize, so hemangiopericytoma should be accepted as potentially malignant.[7] Our case was evaluated as benign because the diameter of the tumor was small and there was no mitosis or necrosis.

Clinical presentation is nonspecific in the form of a slowly growing mass in the head and neck region without pain. Pain may develop if there is nerve compression and when the tumor is localized within the sinus. Other manifestations include telangiectasia, a pulsative mass, warmth of the skin, and a bruit.[6] These signs may be confused with those of vascular malformations or aneurysms. Hypoglycemia, hypotension, secondary hypokalemia due to hyperreninemia, and arteriovenous shunt-induced cardiac failure may develop systemically.[6] In our case, there was a preauricular mass without pain or telangiectasia, pulsation, bruit, warmth, or other systemic signs.

Radiological techniques are used for preoperative evaluation of the tumor. Hemangiopericytoma in the head and neck region has an evident CT appearance of bone remodelling, expansion, and homogeneity.[2] Hypervascularity, septations, and calcifications can be seen in CT scans as well as magnetic resonance imaging (MRI). However, these radiological findings can be seen with pleomorphic adenoma and mucoepidermoid carcinoma, and are thus not specific to hemangiopericytoma.[2] Information on MRI findings of hemangiopericytoma in the head and neck region is not sufficient. Hemangiopericytomas detected in other regions are of high signal intensity on T_2-weighted images and medium signal intensity on T_1-weighted images.[2] In the presence of nonspecific MRI findings, preoperative evaluation of tumor size, localization, determination of vascularity, and relationship with surrounding tissues are very important.

Extensive surgical excision is the treatment of choice. Surgical enucleation should be avoided because the tumor cells may exist outside the capsule. Surgical resection can be performed easily if afferent vessels are bound before the operation.[7] Hemangiopericytomas are radioresistant tumors. Palliative radiotherapy can be used for locally relapsed, inoperable, or incompletely resected cases.[7] Satisfactory results were reported with preoperative embolization, extensive surgical resection, and radiotherapy.[7] The role of chemotherapy is controversial. Some improvement was reported with adriamycin alone or in combination with cyclophosphamide, vincristine, methotrexate, and actinomycin D. However, chemotherapy is not recommended as the first choice treatment, but it can be used in inoperable patients.[8] In our case, the tumor was resected by superficial parotidectomy.

The use of antiangiogenic effect of interferon-alpha was reported in unresectable or metastatic patients, resulting in stabilization in a patient with liver metastasis, and partial improvement in patients with lung metastasis.[9] These findings favor the beneficial effect of adjuvant therapy in the future.

In conclusion, hemangiopericytoma of the parotid gland should be treated with extensive surgical resection because of the risk for local recurrence. Lifelong follow-up is recommended because of the possibility of recurrence and distant metastasis even after long years.

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