

# A rare pathology of the parotid gland: epitheloid hemangioendothelioma

## Abstract

Epitheloid hemangioendothelioma is a tumor of vascular origin with the proliferation of endothelial cells in Epitheloid morphology which is reported very rarely in parotid gland. Clinically it has a borderline acting pattern between hemangioma and angio sarcoma with its local aggressive and metastatic features. World Health Organization put the Epitheloid subtype of hemangioendothelioma to the malign category in soft tissue tumors classification in 2013. Herein we report a case of Epitheloid hemangioendothelioma who had experienced total parathyroidectomy due to a parotid mass in our clinic. In 2years of follow up of this case any recurrences is not determined.

**Keywords:** epitheloid hemangioendothelioma, parotid gland, head and neck malignancy, surgery

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**Abbreviations:** HE, hemangio endothelioma; EHE, epitheloid hemangio endothelioma; CT, computed tomography; US, ultra sound

## Introduction

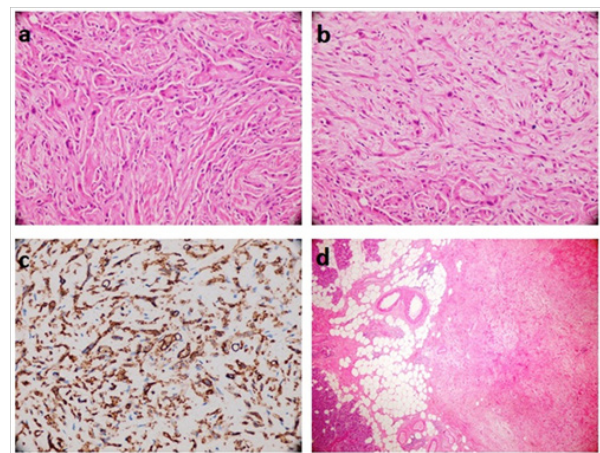
The term Hemangioendothelioma (HE) defines the vascular tumors acting in an intermediate pattern between hemangioma and angio sarcoma. There are different subtypes of hemangioendothelioma with different acting patterns classified as Epitheloid hemangioendothelioma (EHE, malign), capos form HE (intermediate, locally aggressive), ret form HE (intermediate, rarely metastasizing), composite HE (intermediate, rarely metastasizing) and pseudomyogenic HE (intermediate, rarely metastasizing). The most aggressive one among those is the EHE with the highest association rates with metastasis and death due to tumor.

The metastasis risk of EHE is higher than the others. EHE may have both distant and lymph node metastases.<sup>1-4</sup> Although it may be reported in many parts of the body, it is very rarely described in head and neck region especially in major salivary glands. In literature, there are very few number of EHE cases with parotid gland involvement.<sup>5</sup> In this paper, we report a 65years old female patient who has experienced total parathyroidectomy due to a mass in parotid gland and diagnosed with EHE owing to the pathology report.

## Case report

Sixty five years old female patient admitted to our clinic with a progressing mass on the back of left chin for 5months. In head and neck examination, a hard, fixed, painless mass with a dimension of 2x1cm was palpated on posterior of left angle of mandible. Her other otolaryngologic examinations were normal. In neck ultrasound (US), on lateral part of the left parotid gland a heterogeneous, hypoechoic solid mass of 17x11mm was observed. Computed tomography (CT) of the neck revealed a nodular mass of 17x11mm with regular borders on inferior part of the left parotid gland, there was no pathologic lymph nodes reported. Fine needle aspiration cytology was reported as 'suspicious for malignancy'. Owing to this result, left

total parathyroidectomy was performed to the patient under general anesthesia. During operation, a mass in inferior part of the parotid gland was observed and in frozen section malign-benign differentiation of the mass could not be done. Any complications in intraoperative or postoperative periods were not observed. The pathology report of the specimen revealed 'Epitheloid hemangioendothelioma' in postoperative period (Figure 1). Poster anterior lung X-Ray graph and abdominal ultrasound of the patient were normal. In postoperative 2<sup>nd</sup> year control of the patient, any sign of recurrence was not determined.



**Figure 1** (a-b) Anastomosing vascular structures in patients' mass (HE x 200), c) Strongly positive CD-34 immunohistochemistry staining (magnification x 400), d) Primitive vascular differentiation of neoplastic cells with cytoplasmic vacuolization (HE x 100).

## Discussion

Epitheloid hemangioendothelioma is a tumor of vascular origin with the proliferation of endothelial cells in epitheloid morphology which is reported first by Weiss & Enzinger.<sup>6</sup> EHE is generally reported in liver, lung, bone, soft tissue and skin. Involvement of

multifocal areas in these organs can be seen. EHE is reported very rarely in head and neck region and to the best of our knowledge, very few number of EHE cases located in parotid gland are reported to date.<sup>5</sup> It can be seen in every age group but not in childhood. However, it is common after 2<sup>nd</sup> decade. A slight female dominance is present.

Although symptoms can be different according to the location, they are generally nonspecific. They can be presented as painful masses in soft tissue.<sup>2,3</sup> Pigadas et al.,<sup>5</sup> reported a female patient presented with a painful mass in parotid with 1cm diameter.<sup>5</sup> They emphasized that in differential diagnosis parotid gland pathologies including adenoid cystic cancer, infarcted or infected benign lesions, muco epidermoid cancer, lymphoma or a clinic cell carcinoma that can present with pain should be regarded. Our female patient admitted to the clinic with a progressing, painless, solid mass located on the inferior part of parotid gland for 5months.

EHE is regarded as malign tumors owing to its more distinct metastasis rates than other HE types and compared with angiosarcoma.<sup>1</sup> Histopathological they are angiocentric malign vascular neoplasia produced from endothelial cells with Epitheloid morphology in an evident mix-hyaline stroma. Cells with eosinophilic cytoplasm (blister cell) containing vacuoles or erythrocytes with vacuoles are characteristics. Low nuclear graded cases are reported as well as the cases with high graded a typia. Moreover, focal spindle feature and necrosis are associated with worse prognosis. On the other hand recurrences in cases with benign histopathological characteristics are also reported.<sup>1-3</sup>

In recent years, EHE is started to be classified into 2 groups according to tumor dimensions and mitotic activity. Tumor dimension of larger than 3cm and presence of 3 mitoses/50 high power fields are regarded as high risk. Absence of these features is considered as the low risk. Metastasis rates are obviously high in high risk EHE group.<sup>1,7</sup> In EHE, typical vascular markers including CD 34, CD31, FLI 1, ERG and transcription factors are generally determined as positive. The epithelial cell markers of CK7-8-18 and EMA may be positive in 25-40% of cases.<sup>2,4</sup> In this case the differentiation of carcinoma is important. In our case, the dimension of tumor was lower than 3cm and it had low mitotic activity. Moreover in our case, CD34 was positive and Ki- 67 proliferative index was 5-10%.

The diagnosis of EHE in parotid gland is enormously difficult. In preoperative period, clinical, radiologic and fine needle aspiration biopsy findings are not still enough. Although EHE is endothelial in origin, since it is relatively avascular in structure, Doppler ultrasound and angiography cannot be helpful in diagnosis of this tumor.<sup>5</sup> Diagnosis is generally achieved with the histopathological investigation of parathyroidectomy (superficial or total) specimen.

Many EHEs of soft tissue origin have an indolent course. The prognosis of EHE shows obvious differences. In about 1/3 of EHE cases, regional or distant metastases are reported while death due to this disease is seen in 15% of cases. About half of the metastases are reported in regional lymph nodes while the other half are described in distant organs especially in lung, liver and bone. Though the mortality rates of this disease differ according to the involved regions, these rates generally range between 13-43%.<sup>3,4</sup> This course is controversial to the clinical course of many angio sarcomas.

The primary treatment of EHE is the surgery. Especially in patients with low risk, total excision with adequate safety margins around is recommended. On the other hand, in high risk groups, regional lymph node excision should also be added to the surgery. Through these interventions, it should be kept in mind that EHE may be multifocal.

Owing to the slow growing pattern of the tumor, radiotherapy has a limited affect.<sup>1-5</sup> Although some chemotherapy protocols have been tried for EHE, very few have been determined as successful.<sup>7,8</sup> In patients with recurrences, inadequate excision, distant metastasis or high risk, chemotherapy and radiotherapy may be performed. Since tumor dimensions were low, mitotic activity was low, regional lymph node or distant metastasis were absent in our case any additional therapy other than total parathyroidectomy was not performed. Our patient, who did not have any sign of recurrences in 2<sup>nd</sup> year control, is still under follow-up.

## Conclusion

In conclusion, in this paper, we have presented an Epitheloid hemangioendothelioma case in low risk group who was treated successfully and still under follow-up. EHE is a tumor with difficulties in preoperative and perioperative investigations. Heterogeneous stroma and distribution of Epitheloid cells in our case especially made the intraoperative evaluations difficult and malign-benign differentiation could not be made. Moreover, especially in cases expressing endothelial markers in paraffin sections, difficulties in differential diagnosis with unwell-differentiated carcinoma and other sarcomas with Epitheloid morphology are present. Owing to the high mortality risk with metastasis and recurrence risks, EHE should be treated promptly and long follow-up times are recommended. It is a disease that should be kept in mind by the clinicians in differential diagnosis of painful parotid masses though pain was not present in our case.

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## Conflicts of interest

Author declares there are no conflicts of interest.

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## References

- Goldblum JR, Folpe AL, Weiss SW. Hemangioendothelioma: Vascular Tumors of Intermediate Malignancy. (6th edn), In: Goldblum JR, et al. (Eds.), Enzinger and Weiss's Soft Tissue Tumors. (Chapter 22) Saunders. USA. 2014. p.681-702.
- Requena L, Kutzner H. Hemangioendothelioma. *Semin Diagn Pathol*. 2013;30(1):29-44.
- Ravi V, Patel S. Vascular sarcomas. *Curr Oncol Rep*. 2013;15(4):347-355.
- Weiss SW, Antonescu CR, Bridge JA, et al. Epitheloid hemangioendothelioma. In: Fletcher CDM, et al. (Eds.), In World Health Organization Classification of Tumours of soft tissue and bone. IARC press. 2013. p.155-156.
- Pigadas N, Mohamid W, McDermott P. Epitheloid hemangioendothelioma of the parotid salivary gland. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2010;89(6):730-738.
- Weiss SW, Enzinger FM. Epitheloid hemangioendothelioma: a vascular tumor often mistaken for a carcinoma. *Cancer*. 1982;50(5):970-981.
- Deyrup AT, Tighiouart M, Montag AG, et al. Epitheloid hemangioendothelioma of soft tissue: a proposal for risk stratification based on 49 cases. *Am J Surg Pathol*. 2008;32(6):924-927.
- Ye B, Li W, Feng J, et al. Treatment of pulmonary Epitheloid hemangioendothelioma with combination chemotherapy: Report of three cases and review of the literature. *Oncol Lett*. 2013;5(5):1491-1496.