OLGU SUNUMU CASE REPORT

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Pseudomyogenic Hemangioendothelioma of the Retro Auricular Sulcus: A Rare Case of a Vascular Tumor

Retroauriküler Sulkus Yerleşimli Psödomiyojenik Hemanjiyoendotelyoma: Nadir Bir Vasküler Tümör Olgusu

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ABSTRACT Pseudomyogenic hemangioendothelioma (PHE) is a rare neoplasm with vascular origin mostly seen on the extremities. Up to date, the terminology which represents PHE has been a subject to debate. According to the World Health Organization, PHE is classified among the soft tissue tumors. PHE is mostly seen in soft tissue planes and the distal lower extremities are the most affected location of the body. Localization of this tumor on the head and neck region is uncommon. PHE, being a sarcoma does not favor lymphogenic metastasis. As far as we are concerned, this is the first described case of PHE of the external ear and retro auricular sulcus. In this paper, the management of this case and the current treatment options of PHE are discussed.

Keywords: Head and neck neoplasms; neoplasms; connective and soft tissue; soft tissue neoplasms

ÖZET Psödomiyojenik hemanjiyoendotelyoma (PHE), daha çok ekstremitelerde görülen, vasküler kökenli nadir bir neoplazmdır. Günümüze kadar PHE ile ilgili terminoloji literatürde çok defa tartışılmıştır. Dünya Sağlık Örgütüne göre PHE, yumuşak doku tümörleri başlığı altında incelenmektedir. PHE genellikle yumuşak doku planlarında izlenmekle birlikte alt ekstremite distal kısımları PHE'nin en çok rastlandığı lokalizasyonu oluşturmaktadır. Baş-boyun bölgesinde lokalize PHE olguları oldukça nadirdir. PHE bir sarkom olması nedeniyle lenfatik metastazı sık yapmamaktadır. Mevcut literatür incelendiğinde, sunduğumuz vakanın, dış kulak ve retroauriküler sulkusa yerleşmiş, lenfatik metastaz ile birlikte karşımıza çıkan atipik bir PHE vakası olduğu görülmüştür. Bu yazıda, vakanın yönetimi ile birlikte PHE'nin güncel tedavi seçenekleri tartışılmıştır.

Anahtar Kelimeler: Baş ve boyun neoplazileri; neoplaziler; bağ ve yumuşak doku; yumuşak doku neoplazileri

Pseudomyogenic hemangioendothelioma (PHE) is a rare tumor with vascular origin.[1] Localization on the head and neck region is rare. It has a borderline behavior.[2] Distant metastasis or regional lymph node metastasis are not common.[3] Although a fully accepted treatment modality is not present, wide local excision is performed to treat this neoplasm. Local recurrence is one the challenging aspects of PHE. Here in this case report, we present a female patient with a regional lymph node metastasis and discuss the management in light with the current literature.

CASE REPORT

Written informed consent was obtained from the patient who participated in this case. A 71-year-old female patient applied to our clinic with multiple crusted lesions with pain on her left post auricular region and ear lobule (Figure 1). She stated that the le-

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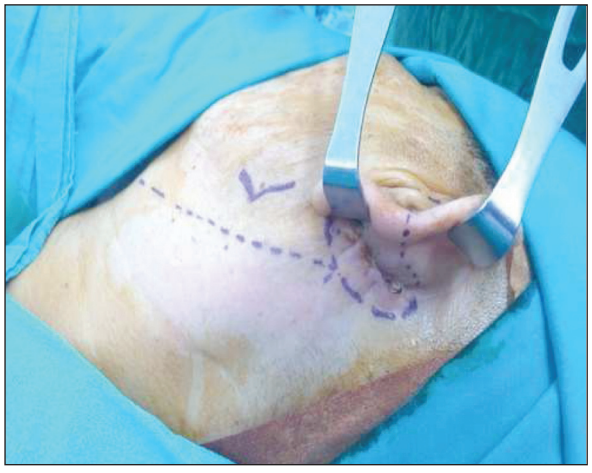

FIGURE 1: The preoperative appearance of the tumor, crusted lesions localized on the retro auricular region is present.

sion first appeared about 6 months ago. The patient was otherwise healthy and without any medication.

On physical examination there was no induration around the lesions though a hyperemic state was present. The patient described a painful sensation during palpation. There was a single palpable lymph node just inferior the parotid tail. There were no additional otorhinolaryngologic findings.

In order to obtain a histopathological diagnosis an incisional biopsy was scheduled. The specimen involved atypical fusiform cells with distinctive nucleoli on a chondromyxoid ground and necrosis (Figure 2). Immunohistochemical studies were performed and the specimen was positive for CD31 (Figure 3). Based on these findings, a diagnosis of PHE was made.

An ultrasonographic evaluation of the neck revealed an intraglandular (3*5 mm) and another lymph node (12*5 mm) located just inferior to the parotid tail. Both lymph nodes demonstrated an echogenic hilus. The computed tomography of the temporal bone was evaluated as normal without bone infiltration of the tumor.

As the tumor showed continuity beyond the surgical margins, in order to make a wide local excision and achieve intact surgical margins, a conclusive surgery was planned. The lesion with its underlying periosteum was excised with 2 cm surgical margins. Additionally, the lower half of the cavum concha and the ear lobule was added to the specimen. The palpated lymph node was also added to the specimen and a frozen section was sent for pathological examination. The frozen section of the suspicious lymph node was evaluated as metastatic (Figure 4). Because of the positive lymph node, a superficial parotidectomy and selective neck dissection were performed (Level 1B, 2, and 3).

The surgical specimen was sent for pathological evaluation. The tumor showed peri-neural invasion. Tumor infiltration on the resected conchal cartilage was noted. Ki-67 proliferation was 20%. All surgical margins were intact.

The patient was discharged on the 7th day of hospitalization. An additional adjuvant radiotherapy was

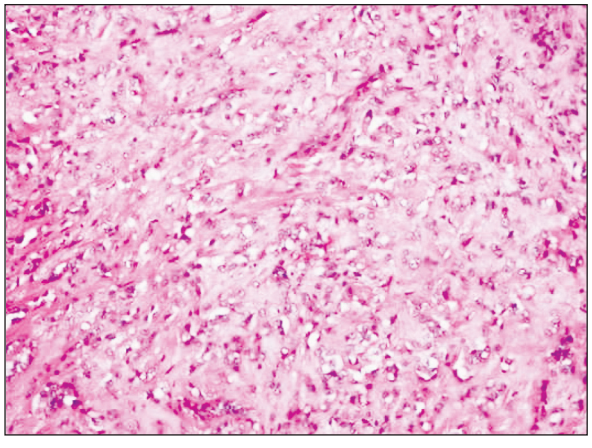

FIGURE 2: Atypical fusiform cells with distinctive nucleoli on a chondromyxoid ground and necrosis are present, x200 magnification, Hematoxylin&Eosin staining.

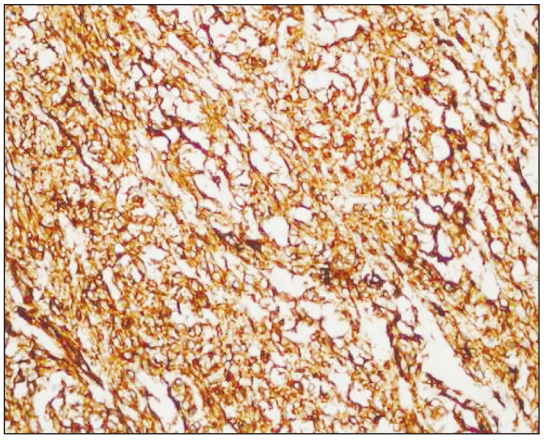

FIGURE 3: CD31 positive staining of the specimen, x200 magnification, Immunostaining.

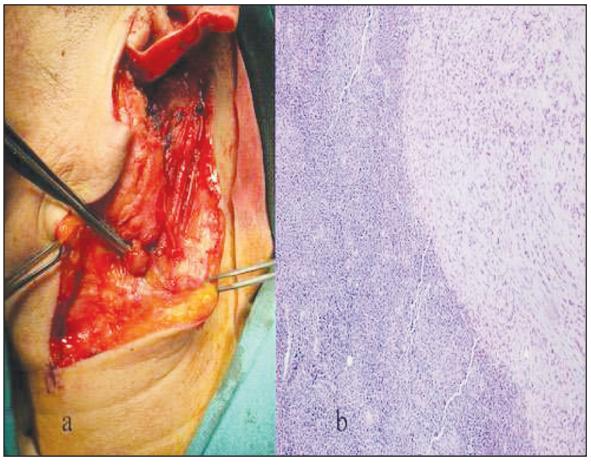

FIGURE 4: a) Intraoperative appearance of the metastatic lymph node; **b)** Microscopic appearance of the metastatic lymph node, x100 magnification, Hematoxylin&Eosin staining

administered. No recurrence was detected on follow ups (1 year). Informed consent was obtained from the patient for this article.

DISCUSSION

PHE is a rare soft tissue tumor with vascular origin. The terminology to represent this tumor has evolved throughout the past 30 years. In 1992, Mirra et al. published an article that pointed out 5 histologically very similar cases that was than misdiagnosed. The retrospective analysis of these cases revealed a fibroma like arrangement of fibro histiocytic and myoid cells therefore they defined this pathology as the fibroma like variant of epithelioid sarcoma.[1] These 5 cases are believed to be the first recognition of PHE. In 2003, Billings et al. published a case series of 7 patients with similar histological findings and by using immunohistochemical stains they have demonstrated the vascular differentiation which than led them to name the tumor as epithelioid hemangioendothelioma.[2] Hornick et al., in 2011, published the largest case series to date and they were the first to use the term PHE.[3] PHE is currently the term that is used in World Health Organization classification of soft tissue tumors.[4]

PHE is mostly seen in the 3rd and 4th decades of life with a distinct male dominancy.[5] Arising from the soft tissue planes, the distal lower extremities are the most affected location of the body. Other anatomic regions that the tumor favors are the upper extremities and the trunk respectively. Sites as the penis, thoracic spine, scalp and oral cavity are only reported in single case reports.[3,6-8] There are also primary cases of bone involvement in the literature.[9] The clinical presentation is mostly with a subcutaneous and nodular mass. Multifocality is not rare. These masses in some cases may be seriously painful while in some, they are asymptomatic. The tumor size is usually not larger than 3 cm.[10] Primary cases of bone involvement usually are with multiple, lytic lesions without periosteal reaction.[9]

The presented case was a 71-year-old female with an unusual localization of PHE. The head and neck region are not one of the areas that PHE is familiar to be seen. There are cases with the involvement of the scalp and oral cavity. Up to date, there is no published data of PHE localized on the auricula or retro auricular sulcus. Osseous involvement is one of the well-known features of PHE however, in our case there was no osseous involvement. However, tumoral infiltration in the resected conchal cartilage was demonstrated.

Histologic appearance of the tumor demonstrates irregular borders with an infiltrative pattern. Plumb spindle or epithelioid cells with vesicular nuclei and a bright eosinophilic cytoplasm form tumor. Mitotic activity is low and there is only mild atypia.[3]

Immunohistochemistry, is how the precise differential diagnosis is made with. PHE demonstrates a diffuse reactivity to cytokeratin AE1/AE3.[2] The tumor is positive for CD31(50%) and negative for CD34.[10] The endothelial transcription factors; FLI-1 and ERG are also positive markers.[3] The genetics of PHE is still a subject that is being investigated. Trombetta et al. managed to manifest a balanced translocation between chromosomes 7 and 19[t (7;19) (q22; q13)].[11]

The first treatment option in PHE is wide local excision. In cases with extensive disease chemotherapy and radiotherapy is preferable. As the molecular biology of the tumor is understood day by day, several different medical treatment options have developed. Targeted therapy with everolimus and sirolimus has initially promising effects.[5,12]

PHE has a relatively indolent prognosis. Local recurrences are common though distant metastasis are rarely reported. Nearly 60% of patients who undergo surgical excision of this tumor experience local recurrences.[5] Up to date, there is only a single patient with regional lymph node metastasis and 3 cases of distant metastasis.[9]

Our case represents the second PHE with a regional lymph node metastasis. Considering the patient's advanced age and positive lymph node in order to prevent another surgical intervention, adjuvant radiotherapy was also administered.

PHE is a rare malignant tumor of vascular origin with a serious risk of local recurrence. This is the first presented case of PHE localized on the auricula and retro auricular sulcus. There is no accepted management guideline of this pathology as the behavior of PHE varies as well. According to our surgical experience, when encountered this lesion, a preoperative imaging of the neck and intra-operative frozen sections from suspected lymph node enlargements must be done. If lymph node metastasis is present, a selective lymph node dissection must be added to the surgical intervention.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Necmi Arslan, Sami Engin Muz;* ***Design:*** *Mustafa İbas;* ***Control/Supervision:*** *Necmi Arslan;* ***Data Collection and/or Processing:*** *Kübra Başarır;* ***Analysis and/or Interpretation:*** *Songül Dursun;* ***Literature Review:*** *Songül Dursun;* ***Writing the Article:*** *Mustafa İbas;* ***Critical Review:*** *Necmi Arslan, Sami Engin Muz;* ***References and Fundings:*** *Songül Dursun;* ***Materials:*** *Kübra Başarır.*

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