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Epithelioid Angiosarcoma of Masticator Space: A Case Report of CT Manifestations and Follow-Up

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Epithelioid angiosarcoma (EA); Masticator space; Computer tomography (CT); Manifestations

1. Abstract

Epithelioid Angiosarcoma (EA) is a special subtype of angiosarcoma, which is common in skin and connective tissue, and its incidence reaches the peak in men aging over 60 years old. EA occurring in masticator space was very rarely reported. We reported EA in a young woman with a lesion located in the right masticator space. Plain Computed Tomography (CT) scan showed irregular blocky soft tissue density shadow with unclear boundary. After enhancement, the lesions showed obviously uneven enhancement in the arterial phase, while the lesions showed uneven decreased enhancement in the venous phase, and there was necrotic tissue with no enhancement. The mass in the right masticator space was removed by extended resection under navigation, and EA was confirmed by postoperative pathology. The tumor recurred 9 months after surgery, in which the tumor in the right masticator space was resected again, and the lesion was completely resected. After surgery, the patient received 30 times of adjuvant radiotherapy and 4 times of chemotherapy. Lung metastasis occurred after 15 months of follow-up, and thoracoscopic radical resection of lung tumor was performed. After 18 months of follow-up, the patient died of multiple organ failure.

2. Introduction

Angiosarcoma (AS) is a rare soft tissue tumor, which mainly occurs in the skin and superficial soft tissues, and it can also affect the deep soft tissues of the limbs, breast, liver, spleen, bone, and other organs. The male to female ratio is about 2:1. Its prognosis is poor, and it is mainly associated with local recurrence and early metastasis [1]. Epithelioid angiosarcoma (EA) is a specific subtype of AS [2]. EA occurring in the masticator space is very rare, and it has been scarcely reported in the literature. We reported the computed tomography (CT) imaging and follow-up data of a case of EA in the right masticator space, and reviewed the literature.

3. Case Presentation

In May 2018, a 28-year-old female patient was admitted to the First Affiliated Hospital of Lanzhou University (Lanzhou, China), complaining of a right temporal mass for more than 4 months. On May 4, 2018, the mass in the right masticator space was biopsied under the guidance of B-mode ultrasound. Immunohistochemical results: tumor cells ckp (3+), vimentin (3+), glial fibrillary acidic protein (GFAP) (-), S-100 (-), CD34 (3+), FacterVIII (focal positive), SMA (-), Desmin (-), bc1-2 (-), ki67 (40%), CD45 (-), HMB45 (-), Melan-A (-), CD31 (mildly positive), and FIL-1 (2+, nuclear positive). Combined with morphology (Figure 1E) and immunohistochemistry, EA was consistently diagnosed. Contrast-enhanced CT of the head and neck showed an irregular blocky soft tissue density shadow in the right masticator space, and the plain CT value was about 35 Hounsfield unit (HU) (Figure 1A). After enhancement, lesions showed obviously uneven enhancement

(Figures 1B, C), and the bone adjacent to the right zygomatic arch showed no obvious abnormality (Figure 1D).

Subsequently, the patient received surgery in the Peking University School and Hospital of Stomatology (Beijing, China) on June 7, 2018. The name of the surgery was enlarged resection of the mass in the right masticator space under navigation + right zygomatic arch resection + right maxilla partial resection + right mandibular coracoid process partial resection + titanium mesh implantation + left thigh anterolateral flap transfer repair + adjacent flap transfer repair. Postoperative pathology confirmed EA. CT reexamination at 20 days after surgery showed that adipose tissue was filled in the surgical area, and the plain CT value was about -95 HU (Figures 2A, B). The right zygomatic arch was absent, which was replaced by preformed titanium mesh (Figure 2B), and the right maxillary bone was absent locally (Figure 2C).

The second CT examination was performed at 48 days after surgery and the following findings were achieved: multiple gas shadows were observed in the posterolateral lesion of the right maxillary sinus (Figures 3A, B), indicating postoperative infection. The postoperative infection debridement and adjacent flap transfer repair of EA in the right masticator space were performed. During surgery, the dead cavity was found under the titanium mesh and at the posterolateral side of the right maxillary sinus, and the diseased tissue was removed. The patient was recovered well postoperatively.

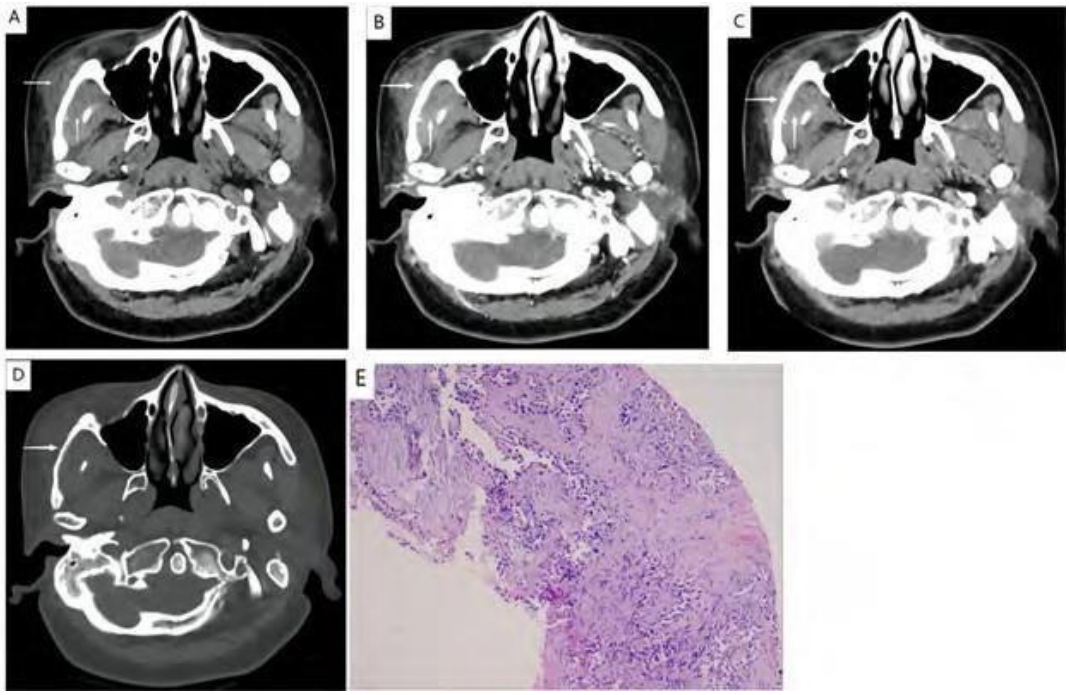

Figure 1: A: Plain CT scan and transverse section of head and neck: soft tissue mass shadows could be observed in the right masticator space (white arrow) and right anterior masticator space (white arrow). The plain CT scan value was about 35 HU, the density was uneven, a strip-like low density shadow could be observed inside, and the boundary was unclear; B: Arterial phase and transverse section of contrast-enhanced CT: the mass (white arrow) was unevenly enhanced, the CT value was about 65 HU, most of the lesions were located in the right masseter muscle, the right temporal muscle was involved, and the border with the right lateral pterygoid muscle was clear; C: Venous phase and transverse view of contrast-enhanced CT: the uneven enhancement of the lesion (white arrow) was weaker than before, with a CT value of about 54 HU. There was no enhancement sign in its internal strip-like low-density shadow, and no clear abnormal enhancement was found in the right lateral and medial pterygoid muscles; D: Head and neck plain CT scan and transverse view, bone window: there was no obvious abnormality in bone of the right zygomatic arch; E: Histological examination, HE staining, and microscopic examination showed that the tumor cells were fat spindle shaped and epithelioid, with large and deep stained nuclei, some nucleoli were visible, the cytoplasm was rich, weakly eosinophilic, cytoplasm was partly bright, and few vacuoles could be observed. Erythrocytes were observed in vacuoles, mitosis was rare, necrosis could be observed locally, followed by interstitial fibrous tissue hyperplasia, partly interstitial hyaline degeneration, tissue congestion, edema, and partly chronic inflammatory cell infiltration (Magnification, 200x).

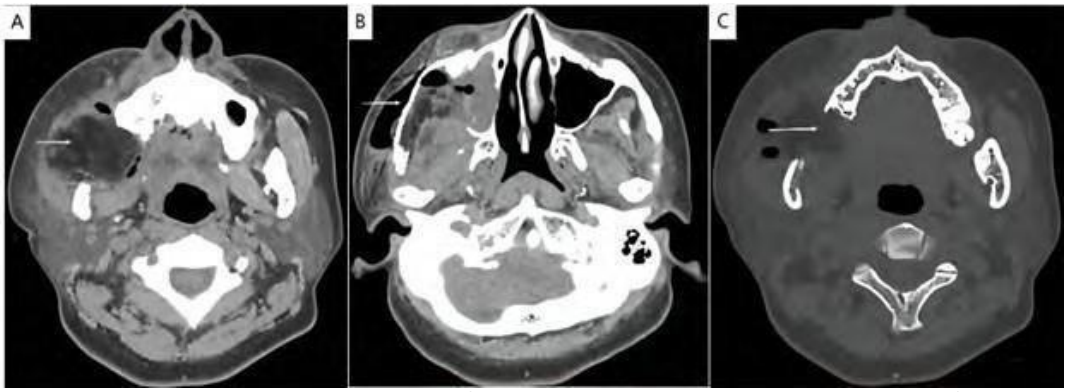

Figure 2: A: Head and neck plain CT scan and transverse view, soft tissue window: very low-density adipose tissue filling could be observed in the right masticator space (white arrow), and the plain scan CT value was about -95HU, with uneven density, strip-like slightly low-density shadow and unclear boundary. B: Plain CT scan and transverse view, soft tissue window: strip-like gas shadow could be observed in the right masticator space (white arrow), and the right zygomatic arch disappeared and was replaced by titanium mesh; C: Plain CT scan, transverse view, bone window: right maxillary bone was absent locally.

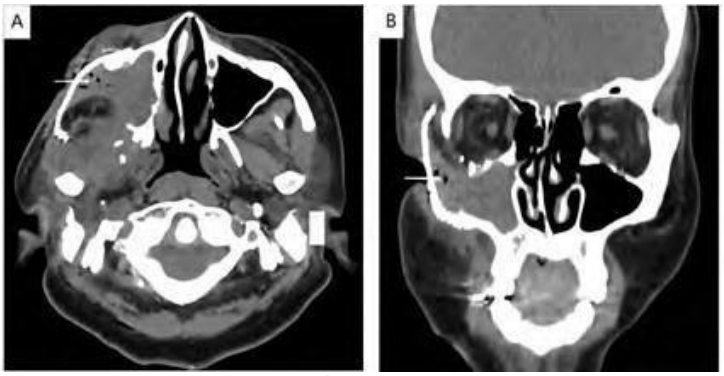

Figure 3: A: Head and neck plain CT scan and transverse view, soft tissue window: very low-density adipose tissue filling could be observed in the surgical area of the right masticator space, and the plain CT scan value was about -95 HU, with uneven density. Large and slightly low-density shadows could be observed in the right maxillary sinus and behind the titanium mesh, the boundary was unclear, and several small round-shaped gas density shadows could be observed inside (white arrows); B: Plain CT scan, coronal view, soft tissue window: large pieces of slightly low-density shadow could be observed in the medial side of the titanium mesh in the right masticator space and in the right maxillary sinus, CT value was about 45 HU, and gas shadow could be observed in the posterolateral side of the right maxillary sinus (white arrow).

The third CT reexamination on March 6, 2019 showed slightly large-scale lower and very low mixed density shadows in the medial side of the titanium mesh in the right masticator space and the posterior part of the right maxillary sinus, and strip-like enhancement was observed in the lesions after enhancement (Figures 4A, B), suggesting recurrence of the tumor after surgery. The following surgeries were performed: titanium mesh removal after EA surgery in the right masticator space + debridement in the right masticator space + mass resection in the right masticator space + right anterolateral thigh flap transfer and repair + right neck vascular exploration + adjacent flap transfer and repair + tracheotomy. During the surgery, necrotic tissue was found under the titanium mesh and in the right maxillary sinus cavity, and the mass was completely removed. The postoperative pathological results showed that there were scattered tumor-like cells in the fibrous tissue, with mild-to-moderate atypia. The recurrence of EA was considered. The surgery was successfully completed and the postoperative recovery was satisfactory.

Since the first surgery, the patient had received 30 times of radiotherapy and 4 times of chemotherapy in the Gansu Cancer Hospital (Lanzhou, China). The specific chemotherapy and radiotherapy regimens were unknown. The fourth reexamination was carried out on September 4, 2019. Chest CT showed solid lobulated nodular shadow in the apicoposterior segment of the upper lobe of left lung, and vacuole sign was observed inside (Figures 5A, B). Head magnetic resonance imaging (MRI) displayed the high-density adipose tissue filling in the operation area of the right masticator space (Figures 5C, D), and the lipid suppression sequence showed the low-density adipose tissue (Figure 5E). No obvious abnormal signal was observed in the operation area in the diffusion-weighted imaging (DWI) sequence (Figure 5F).

The patient underwent general anesthesia, thoracoscopic radical resection of lung tumor, and closed thoracic drainage in Jinchang Integrated Traditional Chinese and Western Medicine Hospital of Gansu Province (Jinchang, China) on September 05, 2019. The postoperative pathological results confirmed EA. After surgery, symptomatic treatments, such as anti-inflammatory and fluid infusion were given, and the patient's clinical status was relieved. After multiple follow-up visits, the patient died at home in December 2019.

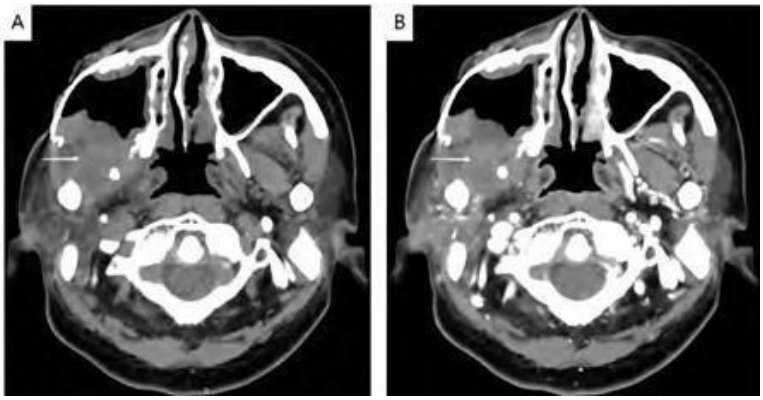

Figure 4: A: Plain CT scan and transverse view of head and neck: small pieces of very low-density adipose tissue could be filled in the surgical area of right masticator space, and the CT value was about –95 HU, with uneven density. On the posterolateral side of the right maxillary sinus and medial side of the titanium mesh, strip-like slightly low-density shadows could be observed, with a CT value of about 52 HU, and the boundary was unclear (white arrow). B: Contrast-enhanced CT and transverse view: the lesions at the posterolateral side of the right maxillary sinus and the medial side of the titanium mesh had an obviously strip-like enhancement, the CT value was about 85 HU, and the boundary was unclear (white arrow).

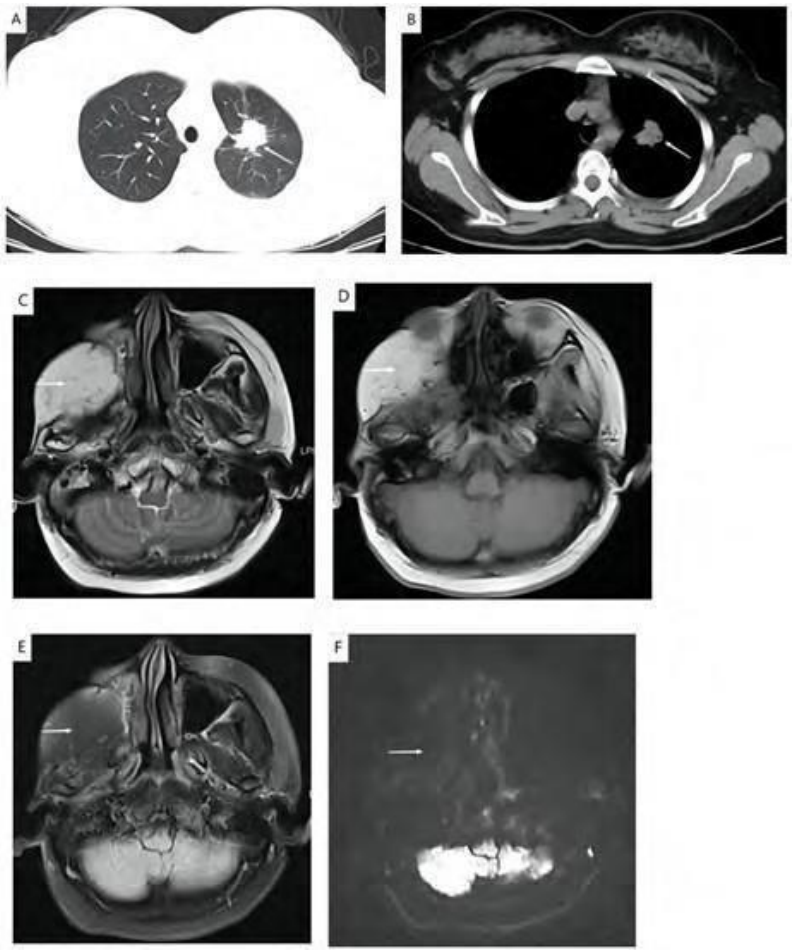

Figure 5: A: Chest plain CT scan and transverse view: solid nodular shadow could be observed in the apicoposterior segment of the upper lobe of left lung, with size of about 2.8×2.6 cm2, CT value was within 27 HU, and with uneven edges, lobulated, and burr sign (white arrows); B: Plain CT scan and transverse section: the density of the lesion was uneven, with vacuole sign (white arrow); C: Head MRI scan, transverse section, T2WI: High-density adipose tissue filling could be observed in the surgical area of right masticator space (white arrow); D: T1WI: High-density adipose tissue filling (white arrow); E: FS: The fat suppression sequence showed a low signal-intensity (white arrow); F: DWI: no obvious abnormal signal (white arrow) was found in the surgical area (white arrow).

4. Discussion

AS is a rare malignant tumor of endothelial origin [3], with an incidence rate of 0.01/100000. The local recurrence rate and metastasis rate of the disease are high. At present, there is no effective treatment plan for AS, and its prognosis is the worst among all soft tissue sarcomas. EA is a special subtype of AS, which is mainly found in the head and face of elderly men, mostly in the superficial part, and only few cases in the deep soft tissue and body cavity were reported [4-6]. EA in the right masticator space of young women was very rarely reported. The etiology and pathogenesis of EA have still remained unknown, which might be related to trauma, radiotherapy, glucocorticoid treatment, and exposure to arsenic [7]. The patient in this case had no history of trauma and no abnormality in her medical history, suggesting that the disease was associated with numerous predisposition factors, the pathogenesis of this disease was complex, and further research is required.

The patient had no specific disease characteristics and imaging manifestations, and had a certain correlation with the location of the disease. Four months before her admission to our hospital, a coin sized tumor was found in the right temporal region of the patient. No treatment was performed, and the mass gradually increased to the size of "broad bean". Specialist examination showed that the mass of about 2×2 cm2 in size could be palpable in the right masticator space, with a hard density and a poor range of motion. It was tender when touched, and the pain could radiate to the eyes and ears. The mouth opening was less than one finger, and the occlusion was normal. CT showed a soft tissue mass in the right masticator space, and the plain CT value was about 35 HU (Figure 1A), with uneven density and unclear boundary. Most of the lesions were located in the right masseter muscle, the right temporalis muscle was involved, and the borders with the right medial and lateral pterygoid muscles were clear. After enhancement, the lesions showed obviously uneven enhancement in the arterial phase, with a CT value of about 65 HU (Figure 1B). The lesions showed less uneven enhancement in the venous phase, with a CT value of about 54 HU (Figure 1C), and there was a strip like low-density shadow without enhancement in the lesions, suggesting necrotic tissue. There was no obvious abnormal enhancement of the right medial and lateral pterygoid muscles. No obvious sign of destruction was found in the surrounding bone (Figure 1D).

Histological manifestations of typical EA could be summarized as follows: It was composed of large atypical malignant cells with hyperchromatic nuclei. Malignant epithelioid cells had abundant cytoplasm, round-shaped nuclei, and prominent nucleoli. Hemorrhage or necrosis was common, and mitotic rate increased [7-8]. It was difficult to make a definite diagnosis based on histopathology alone, and the combination of immunohistochemical techniques was the main method for making a definite diagnosis. The vascular markers of EA included positivity of CD31, CD34, Fli-1, and ERG. CD31 is a specific vascular endothelial marker for diagnosis of AS, while it is typically weakly expressed. CD34 is highly sensitive, while is not specific, and it is positive in 40-100% of cases [9]. Epithelioid sarcoma was found negative for Fli-1, while ERG had a similar positive rate in angiosarcoma and epithelioid tumor [10-11]. Vimentin and CD31 were interstitial and endothelium derived markers. In this case, the two markers, Vimentin and CD31, were positive, indicating that the tumor was originated from interstitial and vascular endothelium. CD34 was mainly existed in precursor cells and vascular endothelial cells derived from blood system. A small proportion of CD34 was positive in this case, which re-confirmed that the tumor was of vascular endothelial origin. The diagnosis should be differentiated from metastasis, epithelioid sarcoma, epithelioid hemangioendothelioma, epithelioid leiomyosarcoma, and other diseases [12].

EA has a low incidence, high invasiveness and poor prognosis, and there is no standard treatment for EA. Extensive surgical resection is the main treatment, which is difficult to completely remove due to its high invasiveness. Postoperative radiotherapy and chemotherapy could be supplemented for EA, while it is still prone to recurrence and widespread systemic metastasis [13]. In this case, the mass in right masticator space was removed by extended resection under navigation and the lesion was basically removed. Postoperative infection debridement was performed 1.5 months after surgery, and the patient was recovered well. The tumor was recurred 9 months after surgery, the mass in right masticator space was resected again, and the lesion was completely resected. After surgery, the patient received radiotherapy for 30 times, and chemotherapy for 4 times. After 15 months of follow-up, lung metastasis occurred and thoracoscopic radical resection of lung tumor was performed. After 18 months of follow-up, the patient died of multiple organ failure. At present, immunotherapy is studying to show anti-tumor activity in the treatment of EA, and its efficacy needs to be further verified [14-16].

5. Conclusions

In conclusion, EA occurring in the masticator space is rare, with the high degree of malignancy, easy to relapse and systemic metastasis, and poor prognosis. The diagnosis is mainly confirmed by immunohistochemistry and assisted by imaging examination. Clinicians, radiologists, and pathologists should deepen their understanding about this disease, so that patients can receive early diagnosis and treatment to improve their prognosis.

6. Acknowledgments

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7. Conflicts of Interest

All authors have completed the ICMJE uniform disclosure form. The authors have no conflicts of interest to declare.

8. Ethical Statement

The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient or legal guardian.

References

1. Erickson LA. Angiosarcoma of the Adrenal Gland. Mayo Clinic Proceedings. 2021; 96: 1376-8.
2. Peng X, Duan Z, Yin H, Dai F, Liu H. Ovarian epithelioid angiosarcoma complicating pregnancy: a case report and review of the literature. J Int Med Res. 2021; 49: 3000605211019641.
3. Bi S, Zhong A, Yin X, Li J, Cen Y, Chen J. Management of Cutaneous Angiosarcoma: an Update Review. Current Treatment Options in Oncology. 2022; 23: 137-54.
4. Hindersin S, Schubert O, Cohnen M, Felsberg J, Schipper J, Hoffmann TK. [Angiosarcoma of the temporal bone]. Laryngorhinootologie. 2008; 87: 345-8.
5. Ai D, Zreik RT, Harris FS, Hill G, Shan Y. Primary epithelioid angiosarcoma of the temporal bone with initial presentation of otalgia. Proc (Bayl Univ Med Cent). 2018; 31: 84-7.
6. Pawlik TM, Paulino AF, McGinn CJ, Baker LH, Cohen DS, Morris JS, Rees R, Sondak VK. Cutaneous angiosarcoma of the scalp: a multidisciplinary approach. Cancer. 2003; 98: 1716-26.
7. Derouane F, Brigitte H, Placide N. Epithelioid angiosarcoma arising after an endovascular aneurysm repair: case report and review of the literature. Acta Clin Belg. 2021; 76: 397-401.
8. Vats K, Al-Nourhji O, Wang H, Wang C. Primary epithelioid angiosarcoma of the mediastinum, cytomorphologic features of a rare entity-A case report and literature review. Diagn Cytopathol. 2022; 50: E181-E7.
9. Martinez C, Lai JK, Ramai D, Facciorusso A, Gao ZH. Cancer registry study of malignant hepatic vascular tumors: hepatic angiosarcomas and hepatic epithelioid hemangioendotheliomas. Cancer Med. 2021; 10: 8883-90.
10. Miettinen M, Wang Z, Sarlomo-Rikala M, Abdullaev Z, Pack SD, Fetsch JF. ERG expression in epithelioid sarcoma: a diagnostic pitfall. Am J Surg Pathol. 2013; 37: 1580-5.
11. Marina M, Corcione L, Serra MF, Ferri T, Silini EM, Ceresini G. Primary Epithelioid Angiosarcoma of the Thyroid in a Patient Occupationally Exposed to Radiations. Front Endocrinol (Lausanne). 2018; 9: 577.
12. Rosenbaum E, Antonescu CR, Smith S, Bradic M, Kashani D, Richards AL, et al. Clinical, genomic, and transcriptomic correlates of response to immune checkpoint blockade-based therapy in a cohort of patients with angiosarcoma treated at a single center. J Immunother Cancer. 2022; 10: e004149.
13. Gao M, Li P, Tan C, Liu J, Tie X, Pang C, Guo Z, Lin Y. Primary Central Nervous System Angiosarcoma. World Neurosurg. 2019; 132: 41-6.
14. Pink D, Andreou D, Bauer S, Brodowicz T, Kasper B, Reichardt P, et al. Treatment of Angiosarcoma with Pazopanib and Paclitaxel: Results of the EVA (Evaluation of Votrient® in Angiosarcoma) Phase II Trial of the German Interdisciplinary Sarcoma Group (GISG-06). Cancers. 2021; 13: 1223.
15. Ren B, Wang W, Tan J, Yuan B, Chen G, Mo X, et al. Efficacy of Anlotinib for the Treatment of Angiosarcoma of the Face and Neck: A Case Report. Front Oncol. 2021; 11: 596732.
16. Di Battista M, Darling MR, Scrivener E, Stapleford R, Wehrli B, McCord C. Histologic and Immunopathologic Variability in Primary Intraoral Angiosarcoma: A Case Report and Review of the Literature. Head and Neck Pathology. 2020; 14: 1139-48.