Asian Journal of Case Reports in Surgery

7(2): 5-9, 2021; Article no.AJCRS.65141

A Rare Case Report of a Young Adult Male Patient Presenting with Angiosarcoma of the Mucogingival Mandible

Septiani Rizka Vinkan[1*] and Soewoto Widyanti[2]

[1]Faculty of Medicine, Sebelas Maret University, Surakarta, Indonesia.
[2]Department of Surgery, Oncology Division, Faculty of Medicine, Sebelas Maret University, Surakarta, Indonesia.

Authors' contributions

This work was carried out in collaboration between both authors. Both authors designed the study, reported the case, wrote the first draft of the manuscript and managed the literature searches. Both authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Asmaa Fathi Moustafa Hamouda, Jazan University, Saudi Arabia.
Reviewers:
(1) Zachariah Chowdhury, Homi Bhabha Cancer Hospital/MPMMCC, India.
(2) Barosa, Joao, Aveiro's Hospital, Portugal.
(3) Ebru Ipek Turkoglu, Izmir Kemalpasa State Hospital, Turkey.
Complete Peer review History: http://www.sdiarticle4.com/review-history/65141

Case Report

Received 24 November 2020
Accepted 29 January 2021
Published 18 February 2021

ABSTRACT

Aims: To report a rare case of angiosarcoma in young adult patient, the flow of diagnosis, and management of the case.
Case Description: We reported a 36-year-old male came to the Surgical Oncology Division, Department of Surgery, Dr. Moewardi Hospital Surakarta with a lump in the molar region of his right mandible. Intraoral examination revealed a reddish soft humped oval mass with a necrotic portion. Wide excision was performed with histopathological results indicating angiosarcoma and chemoradiotherapy was planned. The patient did not check up because of the Covid-19 pandemic until one month later as the mass grew back quickly and bled easily. The second-wide excision was performed, and the histopathological results of necrotic tissue fragments and extensive bleeding were obtained; pleomorphic spindle cells, low cytoplasm, crude chromatin, and mitotic nuclei tended to be an angiosarcoma confirmed by positive CD31 immunohistochemical result. One week after the second excision, the mass was found to regrow, and the patient had metastases to the lung and liver. The patient died within 3 months of the diagnosis.

**Corresponding author: E-mail: rizkavs@gmail.com;*

Discussion: Intraoral angiosarcoma is extremely rare. Angiosarcoma is aggressive with frequent local recurrence and distant metastases. Its progression is influenced by several factors, including age.
Conclusion: Angiosarcoma is a rare tumor and has a high rate of progression at a young age. Angiosarcoma of the oral soft tissue is very rare. Early diagnosis and prompt and precise treatment are fundamental to increasing survival rates.

Keywords: Angiosarcoma; young adult; progressivity.

1. INTRODUCTION

Angiosarcoma cases contribute to less than 1% of all sarcomas. It can occur de novo (primary angiosarcoma) or be secondary to irradiation or in patients with long-term lymphedema (secondary angiosarcoma) [1,2].

This cancer can occur in a variety of locations, most commonly in skin, breasts, deep soft tissues, visceral organs, and bones, by order [2]. The author reports highly aggressive mandibular mucogingival angiosarcoma in a young adult patient.

2. PRESENTATION OF CASE

A 36-year-old male came to the Oncology Surgery Polyclinic Dr. Moewardi Hospital with a lump on the lower right gum since 3 months before, which grew on the molar, which fell off five years ago. The lump that initially had a size 20 x 10 x 5 mm and grew bigger within a month, was painless, did not bleed easily, and had a raised wound. No signs of distant metastases were found from chest x-ray and abdomen ultrasound. The history of radiation exposure was denied.

Intraoral examination revealed a reddish soft humped oval mass on the mucogingival of the right mandible with a necrotic part of 4 x 3 x 3 cm.

The patient underwent wide excision with the histopathologic result indicating angiosarcoma; with pyogenic granuloma, squamous cell carcinoma, and metastasis tumor were considered as differential diagnosis. Because the tumor was excised in toto (altogether), post operation staging evaluation of the tumor was planned to guide the treatment decision-making. Unfortunately, the patient did not check up because of the Covid-19 pandemic until one month later as the mass grew back quickly and bled easily. The mass size reached 10 x 5 x 4 cm.

Preoperative radiotherapy before the second surgery was not applied to this patient because the queue of this procedure in our facility is quite long due to Covid-19 pandemic situation. Chemotherapy was not performed as well because patient Karnofsky Score's was less than 50%. The second-wide excision was performed with histopathological results of necrotic tissue fragments and extensive bleeding, obtained pleomorphic spindle cells, little cytoplasm, nuclei with coarse chromatin and mitosis, leaning an angiosarcoma, confirmed by positive CD31 immunohistochemical result.

2.1 Outcome and Follow-up

One week after the second-wide excision, the mass was found to regrow. The evaluation results showed the presence of lung and hepatic metastases. The patient died within 3 months of the diagnosis.

3. DISCUSSION

Intraoral angiosarcoma very rarely occurs. Several case reports and case series were found in the literature. Clinically, several lesions can be a differential diagnosis of intraoral angiosarcoma, including pyogenic granuloma, hemangioma, Kaposi's sarcoma, malignant melanoma, and other epithelioid neoplasms [3,4,5].

The final diagnosis of angiosarcoma can only be made based on a combination of the clinical findings with the histologic and immunohisto chemical features. Angiosarcoma has a variety of histopathological presentations, so that immu nohistochemistry studies with von Willebrand factor, CD31, CD34, or factor VIII-related antigen are needed to confirm the diagnosis [6]. In this case, the diagnosis was confirmed by positive immunereactivity with CD31.

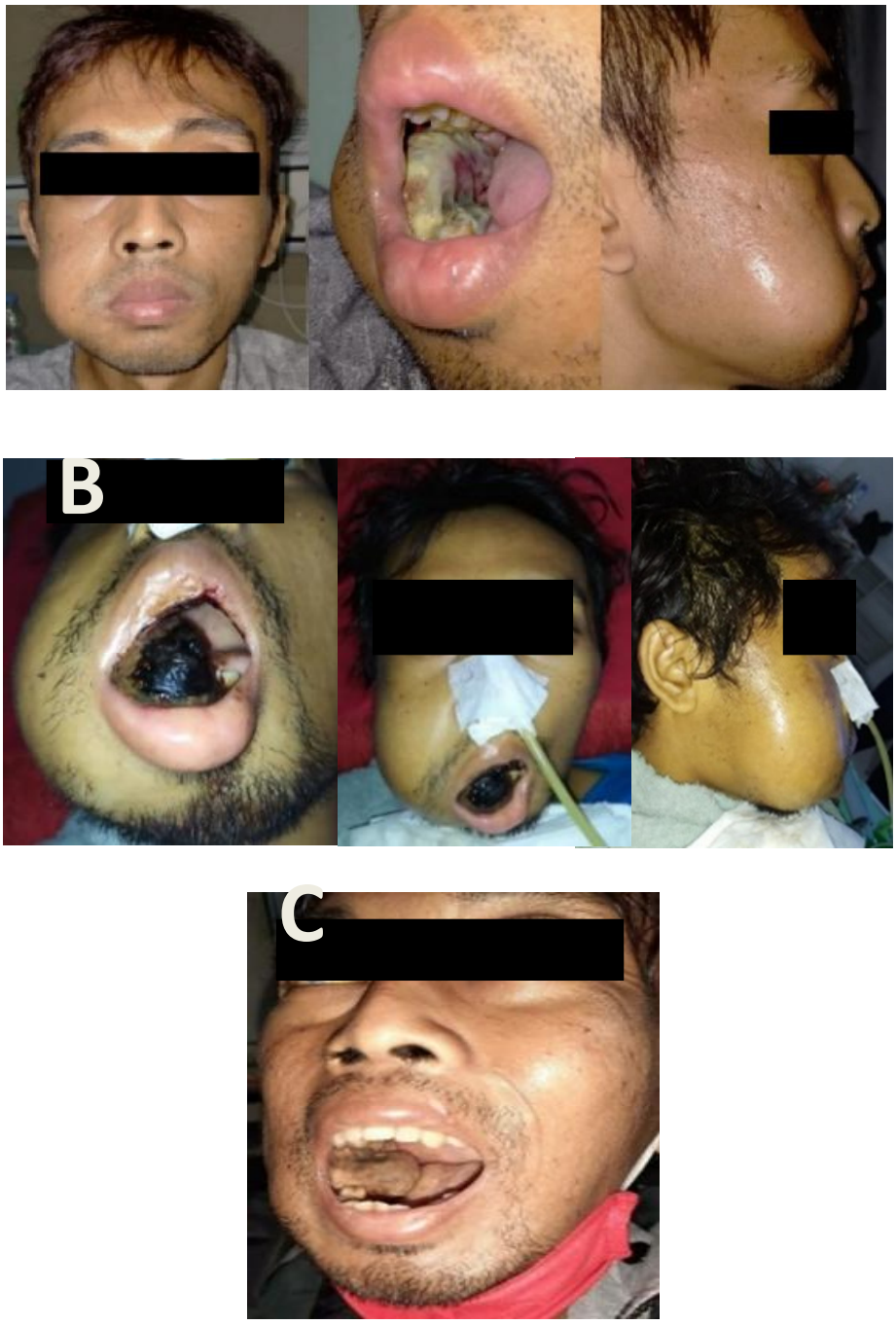

Fig. 1. A. Clinical photo of first visit (preoperative); B. Clinical photo after 3 months (after the first operation); C. Clinical photo of 1 week after the second operation

Fanburgh Smith et al. [3] reported 22 cases of primary angiosarcoma of the oral cavity and salivary glands. Men and women were affected equally. Patient ages ranged from 6 to 90 years (55 years on average). The symptoms include an enlarging and bleeding mass. Histologically, all tumors were vaso formative, 86% had solid areas, and 17% had papillary areas. Immunohistochemically, F-VIII-RA was positive in 19/21 patients, CD31 in 16/19 patients, CD34 in 7/12 patients, and Ulex lectin in 1/1 patients. Survival rates differ by location,

ranging from 1 to 20 years with a mean of 7.3 years [3].

Buehler et al. [7] in their 25-year study of 81 patients showed that negative significant predictors of survival were metastasis at diagnosis, location of tumor in the visceral/deep soft tissue, tumor size greater than 5 cm, tumor necrosis, and non-surgical excision [7].

Patients with metastases had poorer overall survival. Overall survival was also influenced by the primary location, performance status, and the presence of necrosis in the initial tumor sample. Angiosarcoma of the liver, bones, and heart is associated with a poor prognosis [8].

Angiosarcoma mainly affects adults and older people (geriatrics). Deyrup et al. [9] studied 15 cases of angiosarcoma in children and young adults less than 21 years of age. The tumor was mainly located in the heart and mediastinum, with a high mortality rate within 18 months after diagnosis [9,10].

Ferrari et al. [10] studied vascular malignancies in pediatrics, 12 of which were angiosarcoma with a mean patient age of 1-16 years, predominantly male (75%). This suggests that angiosarcoma can also occur in the pediatric population and this diagnosis should be considered in atypical vascular tumors that occur in the mediastinum [9,10].

The treatment of angiosarcoma should be adapted to the staging. However, no prospective randomized study can be a standard therapy because of its rare presentation. Option of treatment of angiosarcoma are wide excision, neck dissection (considered for patients with metastasis to cervical lymph nodes), radiation therapy and chemotherapy (as palliative treatment) [11,12,13].

Wide excision (surgical resection with free margins) is a therapeutic option. Nevertheless, in most cases, complete excision is not possible because of the extension and multifocal nature of the disease. Because of the difficult resection with free margins, tumors on the head and neck or greater than 5 cm have the worst prognosis, so early diagnosis is fundamental to improving survival in these patients [11,12,13].

Scott et al. [14] in their 35-year study in University of Florida reported 41 patients with angiosarcoma were treated with radiotherapy. The median patient age was 67 years. 31 patients were treated with both surgery and radiotherapy and 10 patients were treated with only radiotherapy. The patients with the best outcomes were treated with surgery and radiotherapy 3 times daily [14].

To control the bleeding of the tumor, there can be 2 options management, with radiotherapy and surgery. In this case, preoperative radiotherapy before the second surgery was not performed because the queue of this procedure in our facility is quite long. The second-wide excision was decided to controlled the bleeding. We planned chemotherapy to this patient with taxanes or anthracyclines.

In this case, the patient's clinical condition deteriorated rapidly. The patient had difficulty of breath and scleral icterus. Pleural effusion (pulmonary metastases), as well as hepatomegaly and multiple liver nodules (liver metastases), were found. Laboratory results show the transaminase enzyme significantly increased. The results of the echocardiogram evaluation showed an extracardiac mass pressing on the right side of the heart, as well as mitral regurgitation and mild tricuspid regurgitation. The patient died within 3 months of the diagnosis.

Primary tumor in the mediastinum (can occur as atypical vascular tumors) should be taken as possible disease in this case, given the high progression of this patient. Angiosarcoma in mediastinum usually occurs in pediatric population, but this case were occurred at young adult. At our department in Dr. Moewardi Hospital Surakarta, we had found 3 patients with same diagnosis (angiosarcoma of oral cavity) including this case. The 2 other patients are geriatric population and had better survival rate. They had completed the adjuvant chemotherapy with no local recurrence.

4. CONCLUSION

Angiosarcoma has a high rate of progression at a young age. Angiosarcoma of the oral soft tissue is very rare, and is an aggressive neoplasm that results in frequent local recurrence, early metastasis, and a low survival rate. Early diagnosis and prompt and precise treatment are fundamental to increasing survival rates.

CONSENT AND ETHICAL APPROVAL

As per university standard guidelines, participant consent and ethical approval have been collected and preserved by the authors.

ACKNOWLEDGEMENTS

The authors received no specific grants from any funding agency in the public, commercial or not-for-profit sectors

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Tabareau-Delalande F, Muret A, Miquele storena-Standley E, Decouvelaere A, Pinieux G. Case report cutaneous epithet lioid clear cells angiosarcoma in a young woman with congenital lymphedema. Case Reports in Pathology; 2013. Article ID: 931973.
2. Fayette J, Martin E, Piperno-Neumann S, Le Cesne A, Robert C, Bonvalot S et al. Angiosarcomas, a heterogeneous group of sarcomas with specific behavior depending on primary site: A retrospective study of 161 cases. Annals of oncology. European Society for Medical Oncology. 2007;18: 2030–2036.
3. Fanburg-Smith J, Furlong M, Childers E. Oral and salivary gland angiosarcoma: A Clinicopathologic study of 29 cases. The united states and canadian academy of pathology, Inc. 2003;16(3):263.
4. Terada T. Angiosarcoma of the oral cavity. Head and Neck Pathol. 2011;5:67–70.
5. Hartanto F, Lau S. A case report of angio sarcoma of maxillary gingiva: Histopatho logy aspects. Scientific dental journal. 2018;77-83.
6. Aditya A, Lele S: A nodular growth on maxillary gingiva. Indian J Dent Res. 2012; 23(1):116-119.
7. Buehler D, Rice S, Mood J, Rush P, Hafez G, Attia S et al. Angiosarcoma outcomes and prognostic factors: A 25-year single institution experience. Am J Clin Oncol. 2014;37(5):473–479.
8. Jae-Joon Kim, Seyoung Seo, Jeong Eun Kim, Sung-Ho Jung, Si Yeol Song, Jin-Hee Ahn. Clinical outcomes of angiosarcoma: A single institution experience. Cancer Commun. 2019;39:44.
9. Deyrup A, Miettinen M, North P, Khoury J, Tighiouart M, Spunt S, et al. Angiosar comas arising in the viscera and soft tissue of children and young adults a clinicopatho logic study of 15 cases. Am J Surg Pathol 2009;33:264–269.
10. Ferrari A, Casanova M, Bisogno G, et al. Malignant vascular tumors in children and adolescents: A report from the Italian and german soft tissue sarcoma cooperative group. Med Pediatr Oncol. 2002;39:109–114.
11. Rampinellia H, Ramos-e-Silvaa M, Car valho D, Nurimar Q, Fernandesa C. Cuta neous angiosarcoma. Case Rep Dermatol. 2018;10:55–60.
12. Requena C, Sendra E, Llombart B, San martín O, Guillén C, Lavernia J, et al. Cutaneous angiosarcoma: Clinical and pathology study of 16 cases. Actas Dermo sifiliogr. 2017;108:457–465.
13. Grundahl JE, Hallermann C, Schulze HJ, Klein M, Wermker K. Cutaneous angiosar coma of head and neck: A new predictive score for locoregional metastasis. Transl Oncol. 2015;8:169–175.
14. Scott M, Portnow L, Morris C, Marcus RB, Mendenhall NP, Mendenhall WM, Indelicato DJ. Radiation therapy for angio sarcoma: The 35-year University of Florida experience. Am J Clin Oncol. 2013;36(2):174-80.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/65141