A GIANT CARCINOMA EX PLEOMORPHIC ADENOMA OF THE SOFT PALATE

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ABSTRAK
Karsinoma eks adenoma pleomorfik (Ca eks PA) adalah keganasan epitel yang timbul dari adenoma pleomorfik jinak primer atau rekuren pada kelenjar ludah. Penilaian histopatologi merupakan baku emas untuk menegakkan diagnosis. Tujuannya adalah untuk menentukan pengobatan yang tepat mengingat kemiripan gejala klinis Ca eks PA dan adenoma pleomorfik jinak, dengan mempertimbangkan kekhasan tumor ini. Seorang wanita berusia 55 tahun datang dengan massa berbentuk oval di area junctional palatum molle selama 8 bulan. Massa tersebut berbatas tegas dan melekat pada jaringan di sekitarnya. Massa ini menyebabkan gejala disfagia dan sering terjadi rinolalia. Pemeriksaan histopatologi pascaoperasi memastikan diagnosis Karsinoma eks adenoma pleomorfik (CA-eks-PA). Diagnosis yang akurat dan tatalaksana bedah pasien dengan Ca eks PA dapat meningkatkan angka kesintasan mereka.

Kata kunci : karsinoma eks adenoma pleomorfik, palatum, rinolalia

ABSTRACT
Carcinoma ex pleomorphic adenoma (Ca ex PA) is an epithelial malignancy arising from a primary or recurrent benign pleomorphic adenoma in the salivary glands. Histopathologic assessment is the gold standard for making the diagnosis. Objective to determine an appropriate treatment due to the resemblance of clinical symptoms of Ca ex PA and benign pleomorphic adenoma, considering the peculiarity of this tumor. A 55-year-old female presented with an oval shaped mass in the junctional area of the soft palate for 8 months. The mass was well demarcated and adherent to adjacent tissue. This mass led to symptoms of dysphagia and frequent of rhinolalia. Histopathologic examination post-operative confirmed the diagnosis of Carcinoma ex pleomorphic adenoma (CA-ex-PA). An accurate diagnosis and surgical management of patients presenting with Ca ex PA could increase their survival rates.

Keywords *: carcinoma ex pleomorphic adenoma, palate, rhinolalia*

INTRODUCTION

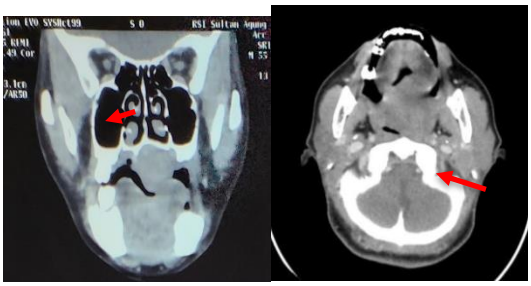
Pleomorphic adenoma (PA) is the most common benign salivary gland tumour. It is mostly found in parotid glands. Around 6% of these tumours transform into Carcinoma ex pleomorphic adenoma. Carcinoma ex pleomorphic adenoma (Ca ex PA) is defined as a carcinoma arising from a primary or recurrent benign pleomorphic adenoma (PA). Ca ex PA predominantly affects the major salivary glands. It has been known to manifest in the minor salivary glands in the oral cavity, especially the hard and soft palate. Most of the Ca ex PA can be asymptomatic, also not widely invasive, and often have similar clinical presentations as PA. Symptom of pain usually results from local extension of the neoplasm into adjacent soft and hard tissues (Sedassari BT, Lascane NA, Tobouti PL *et al*. 2014).

The exact pathogenesis of Ca-ex-PA is not clear. The disease is also uncommon and often poses a diagnostic challenge to clinicians and pathologists. Histopathological examinations is an important aid in diagnosis. An accurate diagnosis and surgical management prevent recurrence. We present here a Carcinoma ex pleomorphic adenoma of the soft palate and appropriate treatment.

CASE REPORT

A 55-year-old female patient was admitted to the Otorhinolaryngology polyclinic of Sultan Agung Islamic Hospital with complaints of a mass on her hard and soft palate since eight months ago, the mass was enlarging slowly and painless. The patient also complained of disturbance of speech and lumpy in her throat which leads to rhinolalia and frequent of dysphagia. From the intra oral examination revealed an oval-shaped mass in the junctional area of soft and hard palate, well-demarcated and adherent to adjacent tissue. On palpation, the lesion was hard and firm in consistency and also painless. The patient had no enlarged of cervical lymph nodes (Figure 1). The paranasal sinus computed tomography scan coronal plane in oropharyngeal centration with contrast showed that there is an isodense lesion with clear borders and regular edges in the palate to the oropharyngeal region, with a size of approximately 5,97x4,62x3,82 centimeters. Post-contrast injection, homogeneous enhancement appears (Figure 2).

Figure 1. The Mass of The Soft Palate On Oral Examination



(A) (B)

Figure 2. The Isodense Lesion in the Palate To The Oropharyngeal Region On Computed Tomography with Contrast ; (A) Coronal View, (B) Axial View

The patient underwent a surgical excision under general anesthesia based on anamnesis, physical examination, and computed tomography examination. During the surgery, the mass of the palate was excised. The tumour was removed entirely (Figure 3). To prevent the stabilization of mucosa soft palate, remaining of the palate mucosa was sutured to adjacent tissue (Figure 4). Tumor tissue was taken to histopathology.

The patient was discharged after three days of hospitalization, and was called up for follow-up in the first week. In the first week follow up the symptoms of disturbance of speech and lumpy on her throat which leads to rhinolalia and frequent of dysphagia were significantly improved. The palate mucosa sutured showed good condition.

Figure 3. Dissected Specimen

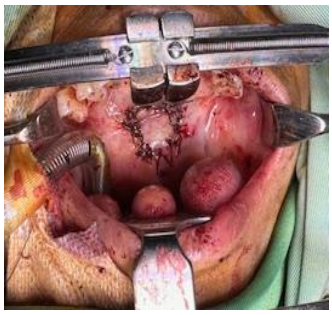


Figure 4. Post Operative View The Palate Mucosa Was Sutured

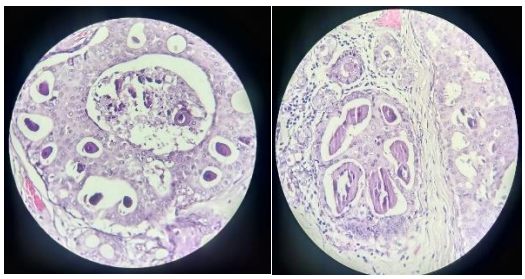


Figure 5. The Histopathology Showed Tubular, Infiltrative, and Trabecular Structures

The histopathology examination was done in the first week of follow-up, with the result showing the mass invades the fibro-collagen and trabecular connective tissue of the bone. Tubular, infiltrative, and trabecular structures (Figure 5). The nuclei tend to be granular, and hyperchromatic, with more pleomorphic mesentery cells in the fibromyxoid and chondroid stroma. The conclusion is in accordance with carcinoma ex pleomorphic adenoma. Based on the histopathology results, the patient was consulted by a Consultant and Specialist of Otorhinolaryngology Oncology to administer chemotherapy concurrently with radiotherapy as an adjuvant therapy. Currently, the patient has passed eight-cycles chemotherapy combination of paclitaxel and cisplatin. During chemotherapy, there are no serious side effects in the patient. The patient will be administered to radiotherapy as an adjuvant therapy and to reduce the risk of cancer recurrence.

DISCUSSION

Carcinoma ex pleomorphic adenoma (Ca ex PA) is a malignant neoplasm arising from a primary or recurrent pleomorphic adenoma (PA) in the salivary glands. This tumor is rare, poorly understood malignancy, and very aggressive. The prevalence of Ca ex PA is 5.6 cases per 100,000 malignant neoplasms and an annual incidence rate of 0.17 tumors per million persons. The tumor is found slightly more common in women than men with ratio of 3:2.[6] The most common site is in the parotid glands. In the minor salivary glands, this tumor frequently originates in the hard and soft palates. The most common clinical presentation of Ca ex PA is

a firm mass and painless. The etiology of Ca ex PA is still uncertain, mostly allegedly because of the involvement of environmental and genetic factors. Some factors of predisposition are genetic, suspected of additional exposure to radiation, users of tobacco, chemicals exposure, and viruses. In this patient, genetic factors and passive exposure from tobacco users might be a risk factor (Farhat, Rizalina A, Asnir *et al*, 2018).

The exact pathogenesis of Ca ex PA is still unclear. Ca ex PA can be asymptomatic as most of the cancers are not widely invasive and often have similar clinical presentations as PA. In the present case, the growth of the hard and soft palate mass in resulted a lumpy on her throat which lead to rhinolalia and frequent of dysphagia for almost eight months. Frequently, patients become aware of the cancer when they experience rapid enlargement of the mass, pain, or other clinical symptoms. On the other hand, patients with Ca ex PA may carry a slow growing mass for before coming to clinical attention. In general, the primary treatment for Ca ex PA is adequate surgical resection of the tumor. Chemotherapy can also be administered concurrently with radiotherapy as an adjuvant therapy or for distant metastasis (Chooback N, Shen Y, Jones M *et al*, 2017).

Livolsi and Perzin stated that lesions of the palate had a better prognosis as compared to tumors of the major salivary glands. Different from their statement, Spiro *et al*. reported that Ca ex PA of the palate a developed high recurrence rate. The completeness of tumor resection contributes to a good prognosis. In this patient, after undergoing surgical excision the histopathology results showed carcinoma ex adenoma pleomorphic. Ca ex PA can be classified as non-invasive Ca ex PA, minimally invasive Ca ex PA, and invasive Ca ex PA, depending on the level of invasion. From histopathology of this patient showed the mass invades the fibrocollagen (Yonekawa AN, Morita Y , Kusuyama Y *et al*, 2002). Because of the extent of invasion was found to correlate with Ca ex PA recurrence and survival, the patient then administered chemotherapy concurrently with radiotherapy as an adjuvant therapy.

CONCLUSION

In conclusion, a proper diagnostic intra-oral examination, computed tomography scan with contrast and histopathology are essential for diagnosis and surgical planning in patients of carcinoma ex adenoma pleomorphic.

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REFERENCES

Antony DJ, Gopalan V, Smith RA *et al*. (2012). *Carcinoma ex Pleomorphic Adenoma: A Comprehensive Review of Clinical, Pathological and Molecular*. *Head and Neck Pathol*. 2012(6):1–9.

Chooback N, Shen Y, Jones M *et al*. (2017). *Carcinoma ex pleomorphic adenoma: case report and options for systemic therapy*. *Curr Oncol*. 2017; 24(3):e251-e254.

Farhat, Rizalina A, Asnir *et al*. (2018). *An Uncommon Occurrence of Pleomorphic Adenoma in the Submandibular Salivary Gland: A Case Report*. *J Med Sci*. 2018; 6(6):1101-1103.

Khanna D , Chaubal T , Bapat *et al*. (2019). *Carcinoma ex pleomorphic adenoma: a case report and review of literature. African Health Sciences*. 2019; 19 (4) : 3253-63.

Kim KM, Lee AH, Yoon SH *et al*. (1997). *Carcinoma ex Pleomorphic Adenoma of the Palate*. *JKMS* 1997 ; 12: 63-6.

Kemenkes RI. (2024). Laporan Prevalensi Hipertensi. https://databoks. katadata.co.id/datapublish/2022/01/25/cakupan-pemberian-asi-ekslusif-di-20-provinsi-ini-masih-di-bawah-nasional

Meidayanti, G., Ayu, S., Candrawati, S. A. K., & Lestari, N. K. Y. (2023). *The Effect of Slow Stroke Back Massage on Blood Pressure in Elderly Patients with Hypertension*. *Holistic Nursing and Health Science*, *6*(1), 30–37.

Nurlathifah, F. A., Cahyani, R., Nugraha, R. M., & Nursiswati. (2022). Efektifitas Terapi *Slow Stroke Back Massage* (Ssbm) Terhadap Penurunan Tekanan Darah Pada Lansia Penderita Hipertensi : *A Systematic Review. Seminar Nasional Keperawatan*, *8*(1), 194–202.

Pramono, J. S., Arsyawina, & Masita, I. K. (2021). *Reducing Blood Pressure with Slow Stroke Back Massage and Warm Water Foot Soak on Isolated Systolic Hypertension Patients. International Journal of Nursing and Health Services (IJNHS)*, *4*(4), 414–422.

Puskesmas Maccini Sawah. (2024). Laporan Data Hipertensi Posbindu dan Puskesmas Keliling Wilayah Kerja Maccini Sawah.

Sedassari BT, Lascane NA, Tobouti PL *et al*. (2014). *Carcinoma Ex Pleomorphic Adenoma of the Palate Composed of Invasive Micropapillary Salivary Duct Carcinoma and Adenoid Cystic Carcinoma Components An Unusual Case With Immunohistochemical Approach*. *Medicine.* 2014 ; 93 (27) : 1-4.

Seok J, Hyun SJ, Jeong WJ *et al*. (2019). *The Difference in the Clinical Features Between Carcinoma ex Pleomorphic Adenoma and Pleomorphic Adenoma*. *Ear, Nose & Throat Journal*. 2019 ;98(8): 504–509.

Srinivas CV. (2020). *Carcinoma Ex-pleomorphic Adenoma: A Case for Cure*. *IJHNS*. 2020; 11(2) : 32-35

Yonekawa AN, Morita Y , Kusuyama Y *et al*. (2002). *Carcinoma ex pleomorphic adenoma in an elderly patient: A case report and literature review*. *Oral and Maxillofacial Surgery Cases*. 2002(8): 100272