

Malakoplakia of the Neck: A Mass Mimicking a Malignancy on Positron Emission Tomography/Computed Tomography

Boyunda Malakoplaki: Pozitron Emisyon Tomografi/Bilgisayarlı Tomografide Maligniteyi Taklit Eden Bir Kitle

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ABSTRACT In the head-neck region, experiencing infectious, inflammatory, and malignant masses is a common fact. However, some pathologies are rarely seen in the head and neck area, and one of them is malakoplakia. Malakoplakia is generally a slow-growing benign tumor in the urogenital region. Diagnosis is made microscopically with the appearance of Michaelis-Guttman bodies. This case report presents a 32-year-old patient with a history of early-stage tongue squamous cell cancer. In the routine follow-up of the patient, a mass with similar features to malignancy and a definitive pathological diagnosis of malakoplakia was detected. In this case report, the characteristics of the patient and the mass are discussed comprehensively.

ÖZET Baş boyun bölgesinde enfeksiyöz, inflamatuvar ve malign kitleler sıklıkla görülmektedir. Ancak bazı patolojiler baş boyun bölgesinde çok nadir görülür. Malakoplaki, genellikle ürogenital bölgede yavaş büyüyen iyi huylu bir tümördür. Michaelis-Guttman cisimlerinin görünümü ile mikroskopik olarak tanı konur. Bu vakada, erken evre dil skuamöz hücreli kanser öyküsü olan 32 yaşındaki bir hasta sunulmaktadır. Hastanın rutin takiplerinde malignite ile benzer özellikler gösteren, kesin patolojik tanısı malakoplaki olan bir kitle saptanmıştır. Bu olgu sunumunda hastanın ve kitlenin özellikleri kapsamlı şekilde tartışılmıştır.

Keywords: Malakoplakia; head and neck neoplasms; positron emission tomography/computed tomography

Anahtar Kelimeler: Malakoplaki; baş ve boyun neoplazileri; pozitron emisyon tomografisi/bilgisayarlı tomografi

Malakoplakia is a chronic, inflammatory benign disease whose etiology cannot be presented clearly. Michaelis and Gutmann first diagnosed this disease in 1902 in the bladder, and Von Hasemann gave the disease its present name in 1903. The name “Malakoplakia” originates from the two Ancient Greek words “malacos” which means soft, and “placos” which means plaque.¹

Although it is generally a disease seen in the genitourinary system, it is also less frequently seen in the gastrointestinal system, retroperitoneal area, lungs, bones, and mesenteric lymph nodes. Head and

neck involvement was diagnosed in 49 patients in a compilation dated 2019. Among the involvement areas in the head and neck region of these patients are successively; skin, tongue, temporal bone and tympanum, larynx and trachea, nose and the sinuses, thyroid, vallecula, and epiglottis, tonsils, submandibular gland, and parotid gland.¹

In this study, a malakoplakia case which mimicked malignancy in the submental region in a 32-year-old female patient after tongue cancer in the follow-up period is presented.

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CASE REPORT

A 32-year-old female patient who has no diagnosed chronic disease and no smoking history was treated by us for tongue squamous cell cancer a year ago by partial glossectomy and unilateral supraomohyoid neck dissection. The postoperative disease stage was pT1N0M0.

Following this result, the patient's follow-up was carried out in regular intervals. In her post-operation examinations on the first and 3rd months, relapse and any pathological findings were not observed. The patient did not have any complaints during her examinations after the operation. In her routine check-up at the 9th month after the operation, there was a 2*2 cm palpable, hard, moving soft tissue in the submental region, which did not give pain and displayed sensitivity in palpation, which was interpreted as lymphadenopathy (LAP). In ultrasonographic imaging, a lymph node of 15*8 mm dimensions in the submental region with cortical thickness and millimetric echogenicity in the cortex was monitored. Furthermore, a few non-pathological lymph nodes smaller than 1 cm were reported, and magnetic resonance imaging (MRI) was suggested for the further analysis.

In the radiological interpretation of the MRI imaging, circular lymph nodes were observed as follows: Round lymph nodes, the largest of which reached 11 mm in submental and 13 mm in left level 2B (Figure 1).

Positron emission tomography/computed tomography (PET/CT) result was interpreted as hypermetabolic LAP (12 mm, SUV_{max} : 5.9) on the neck neighboring the base of the tongue, a biopsy was planned with the suspicion of relapse (Figure 2). Fine needle aspiration biopsy result was non diagnostic. The MRI and PET/CT scan findings were conflicting and was pointing the tonsillary area and anterior serivical region as suspicious, respectively. So we decided to perform panendoscopy and open biopsy to rule out malignancy. The suspicious area identified in the imaging in the operation were analyzed under general anesthesia, biopsies were made on the tongue, the palate and the right tonsil and a neck dissection was performed again over the same incision and the suspicious lymph nodes were excised. The excised material was a cream colored nodular tissue of 2x2x0.7 cm dimensions with an irregular surface. A 1 cm diameter cyst was observed on the cross section full of mucoid material.

In the sections, histiocytic proliferation with wide vacuolized cytoplasm, oval circular vesicular nucleus which formed a partially well-demarcated borders within the fibroadipose tissue were observed (Figure 3). The positively stained concentric laminations in periodic acid Schiff (PAS) staining within histiocytes in certain places, "Michaelis-Guttman" bodies were noteworthy (Figure 3, Figure 4). In light of these findings, the pathology examination was interpreted as malakoplakia. Culture and microbiological samples were not taken before or during the operation, since malignancy was thought to be in the

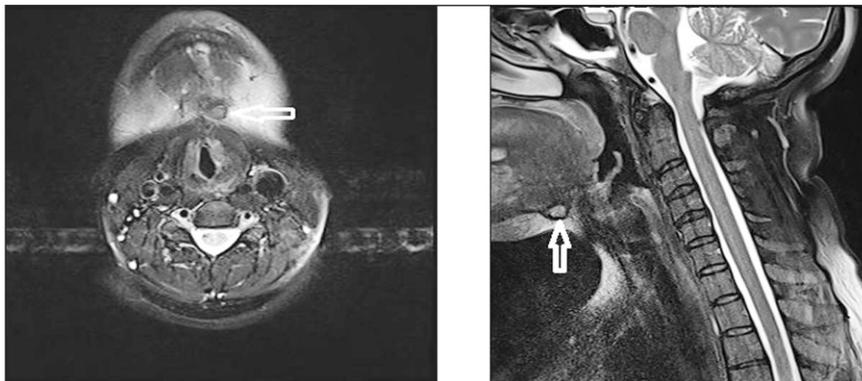


FIGURE 1: T2 image of the mass in axial (left) and sagittal (right) sections. It is noteworthy that it is close to the tongue root in the submandibular area. The tip of the white arrow is malakoplakia.

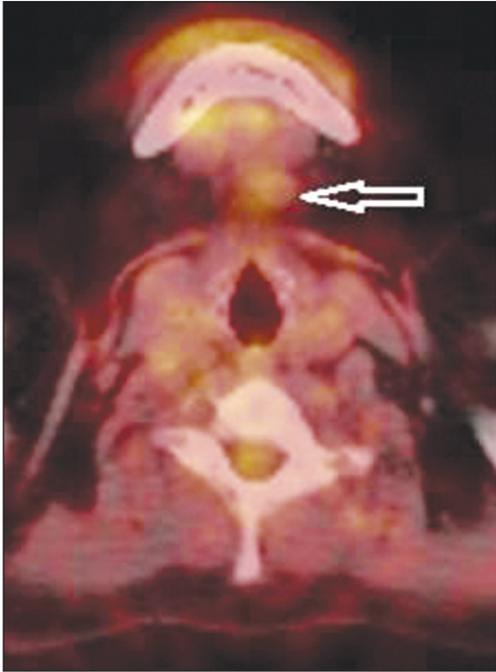


FIGURE 2: At the tip of the white arrow, there is a hypermetabolic mass (12 mm, SUVmax: 5.9) on the neck neighboring the base of the tongue.

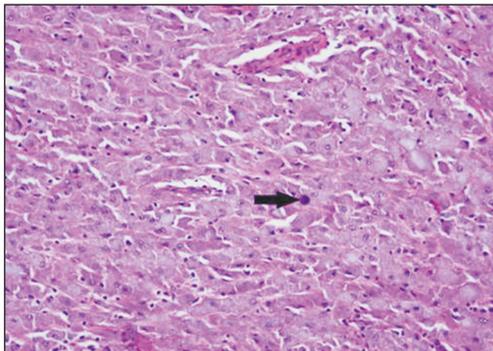


FIGURE 3: Hematoxylin-eosin (HE)x200. Histiocytes and "Michaelis-Guttman" body (black arrow).

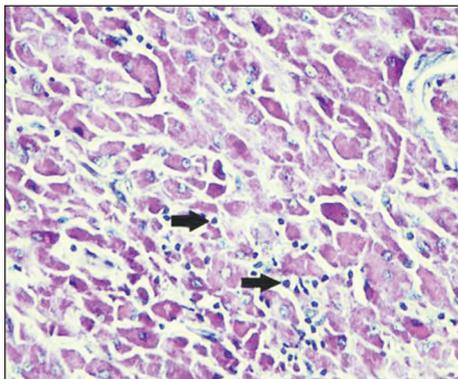


FIGURE 4: Periodic acid-schiff (PAS)x400: "Michaelis-Guttman" bodies (black arrows).

foreground. Therefore, a literature review was made when deciding on antibiotics. Four-week ciprofloxacin 500 mg twice daily treatment, recommended in the literature and frequently preferred in previous similar case series, was started empirically.^{2,3} The patient's long-term follow-ups continue and there is no indication of relapse.

Informed consent was obtained from the patient during this process.

DISCUSSION

Malakoplakia is a benign disease that generally limits itself. It is seen that it can frequently get confused with benign and malignant pathologies.

The age range of the disease is from 6 weeks old to 85 year old, and it is more common in adults compared to children. Distribution of this disease is the same in both genders; however, bladder involvement is more common in women.⁴ The patient presented in this case report is a 32-year-old female patient with a malignancy history.

Abnormally large macrophagic lysosomal granules, which contain a low amount of cyclic guanosine monophosphate (cGMP), play a role in the pathogenesis of malakoplakia. These macrophages cause the reduced beta-glucuronidase release and disrupted bactericidal effect, and this leads to a permanent ineffective inflammatory response and chronic granulomatous disorder.⁵

Clinical appearance and symptoms are usually not specific and depend on the location of the lesion. The most frequently seen symptoms are fever, pain, difficulty in swallowing, sense of a foreign substance in the throat, and reflective otalgia. In the examination, it can be seen in the affected organs of the patient as nodular growth, ulceration, and discharge or regional lymph adenopathy.⁶ In this case report, the patient did not have any symptoms apart from palpable swelling in the submental region and sensitivity.

A histopathological analysis is required for malakoplakia diagnosis. Besides chronic inflammation symptoms, large histiocytes containing periodic acid schiff (PAS) positive granules and circular homocentric structures called "Michaelis-Gutmann"

bodies are observed in the pathological appearance of the disease. If immunohistochemistry tests are applied, the histocytes may be CD68 positive. In this case report, positively stained “Michaelis-Guttman” bodies were observed within the histocytes in PAS staining. Furthermore, positivity was seen in the CD68 and Von Cossa staining.

Inflammatory and infectious conditions such as cancer therapy, foreign bodies, granulomatous processes, and odontogenic abscesses are associated with increased FDG uptake.⁷ An essential component of FDG uptake in tumor tissue is thought to be the involvement of peri-tumoral inflammatory cells, such as macrophages, which demonstrates greater FDG uptake than tumor cells.⁸ In the medical literature, malakoplakia with FDG involvement in the laryngopharynx, pulmonary, and kidneys have been discussed so far.⁹ There are also patients with FDG involvement in the tongue and diagnosed with malakoplakia.^{10,11} A malakoplakia mass, which grows rapidly, can be misdiagnosed as malignancy, as in the case of the patient presented in this report. It can be quite misleading in terms of relapse in patients who have a history of malignancy. The FDG involvement in our patient has been suspicious in terms of malignancy since she had a history of malignancy (SUV_{max} : 5.9). In the literature, the SUV_{max} value in a malakoplakia case in a larynx with FDG involvement has been reported as 14.1.⁹ Increased metabolic activity and the SUV_{max} values reaching the levels seen in malignancy can be misleading. In such cases, carrying out radical procedures before the definitive pathological diagnosis is made, can result in complications.

Numerous treatments have been used successfully from the case reports available in the medical literature, such as; surgical excision, antibiotic therapy, and bethanechol chloride, which theoretically acts to increase cGMP levels.¹² Vitamin C and bethanechol chloride have been used in particular to the patients with weak immune systems as an adjuvant therapeutic strategy in order to increase the function of macrophages.⁶ Combinations of antibiotics involving co-trimoxazole or ciprofloxacin are commonly used due to their excellent intracellular penetration within macrophages.¹³ In malakoplakia treatment, using only

antibiotics results in 50% success. On condition that surgical treatment is performed on its own or in combination with antibiotics, well-nigh 100% success can be achieved.³ In this case, daily 500 mg vitamin C supplements and ciprofloxacin treatment was applied for four weeks as 500 mg twice a day as well.

It should be remembered that malakoplakia can be accompanied by tuberculosis and malignancy in the pathological analysis. Therefore, the tumor mass should be sampled in an extensive manner. In this respect, if there is only malakoplakia in pathology, then the treatment for the disease is anti-biotherapy following up the surgical excision. Specific antibiotics treatment depends on the underlying bacteria indicated in the culture test. In head and neck malakoplakia cases, *Klebsiella*, gram-negative coccobacillus, primarily *Escherichia coli* can be seen.¹

Some lesions in the follow-up period of head and neck cancer may be confusing and may have similar findings of recurrence even cause FDG involvement due to high metabolic activity on PET/CT imaging. Any misdiagnosis in the management of this lesions may lead overtreatment like unnecessary surgical procedures which may have related complications. For this reason, malakoplakia diagnosis should be kept in mind, even if it is seen rarely.

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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

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