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SELÇUK TIP DERGİSİ

Maksiller Sinüste Lokalize Amiloid Tümör

Localized Amyloid Tumor of The Maxillary Sinus

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Özet

Amiloidoma, vücudun tüm organlarını tutabilen, dokularda lokal olarak amiloid birikmesi ile karakterize tümöral oluşumdur. Bununla birlikte baş-boyun bölgesinde ve özellikle de maksiller sinüste oldukça nadir görülen bir hastalıktır. 64 yaşında bayan hasta, yaklaşık 6 aydır mevcut olan burnun sol tarafında tıkanıklık ve kanama şikayeti ile kliniğimize başvurdu. Endoskopik muayenede sol nazal pasajı kapatan sarımtırak görünümde polipoid kitle tespit edildi. Paranazal sinüs tomografisinde sol maksiller sinüsü dolduran ve medial duvarda itilmeye neden olan tümöral kitle tespit edildi. Kitleden daha önce yapılan biopsi sonucu amiloid pozitif olarak rapor edildi. Kesin tanı için kitlenin tamamen çıkarılması önerildi. Hastaya endoskopik medial maksillektomi yapılarak kitle total olarak eksize edildi. Patoloji sonucu amiloidoma olarak bildirildi. Tüm sistemlerin incelenmesi sonucu başka organ tutulumu görülmedi. Rektal biopsisi ve proteinürisi negatif idi. Nadir görülen bu maksiller sinüse lokalize amiloidosis olgusunu sunuyoruz.

Anahtar kelimeler: Amiloidosis, amiloidoma, maksiller sinüs, baş-boyun tümörleri

Abstract

Amyloidoma is a tumoral deposition characterized by localized deposition of amyloid in the tissues. It is very rare in the head and neck region, especially in the maxillary sinus. A 64 years old female patient applied to our clinic with complaints of left-sided nasal obstruction and epistaxis. Endoscopic examination revealed a yellowish polypoid mass obstructing the left nasal passage. Paranasal sinus tomography revealed a tumoral mass filling the left maxillary sinus and repressing the medial wall. Biopsy was positive for amyloidosis. A complete resection of the mass was suggested for a definite diagnosis. The tumor was removed by endoscopic medial maxillectomy, which permitted total excision of the lesion. Pathology results showed amyloidosis. No other organ involvement was detected. Rectal biopsy and proteinuria were negative. Here, we present the extremely rare case having localized amyloidosis of the maxillary sinus.

Key words: Amyloidosis, amyloidoma, maxillary sinus, head and neck tumors.

INTRODUCTION

Amyloidosis is a disease defined by the presence of extracellular deposits of proteinacious material in various tissues. To rule out a systemic amyloidosis is extremely critical as this can markedly change the expected morbidity and mortality. The mean survival of patients with systemic amyloidosis is between 5 to 15 months, whereas patients with localized amyloidosis have excellent prognosis (1). Localized amyloid deposition in the maxillary sinus is an extremely rare phenomenon and a very limited number of cases have been reported so far. The aim of the present article is to present one of such rare cases.

CASE

A sixty four years old female patient applied to our clinic with a left-sided nasal obstruction and intermittent epistaxis. Endoscopic examination revealed a yellowish solid mass in the left nasal cavity. Paranasal sinus tomography demonstrated a presumably tumoral mass lesion filling the left maxillary sinus, causing a scalloping on the medial wall of the sinus and narrowing the left osteomeatal unit and nasal cavity (Fig.1a). In Magnetic resonance imaging (MRI), the 17 mm mass, having a mural contract enhancement in the centre and showing a peripheral contrast enhancement in postcontrast images, was heterogeneous hypodense in T1-weighted images, hyperdense heterogeneous in T2-weighted images, was not supressed in STIR images (Fig.1b). Endoscopic biopsy of the mass was performed and histological examination revealed pink, homogenous amorphous material accumulation, histiocytes surrounding the foci of the amorphous material (Fig.2a). Apple-green birefringence was demonstrated under polarized light with Congo red (Fig.2b). The pathological report was consistent with amyloidosis. Following the pathological report, symptoms suggesting systemic amyloidosis were examined. Moreover, full blood count, renal function tests, full urine tests including Bence-Jones protein, liver function tests, electrocardiogram, chest X-ray, echocardiography, complete abdominal sonography and rectoscopy were performed along with rectal wall biopsy. Examinations revealed no evidence of a systemic amyloidosis. The patient was regarded as having localized maxillary sinus amyloidoma. Then, the patient underwent an endoscopic medial maxillectomy under general anaesthesia and the mass having a 2x1.7x1.7 cm was removed totally. Postoperative period went uncomplicated and the patient was discharged on the postoperative 2nd day. No recurrence was observed during the 6 months follow-up period.

DISCUSSION

Amyloidosis is a clinical disorder having various manifestations based on involvement of different organs. It most commonly affects individuals between 50-70 years of age and occurs predominantly in men. The etiology of amyloidosis is still unclear. It is believed to be a derangement in immunoregulation after protracted antigenic challenge (2). Amyloidosis has 3 forms: primary amyloidosis, secondary

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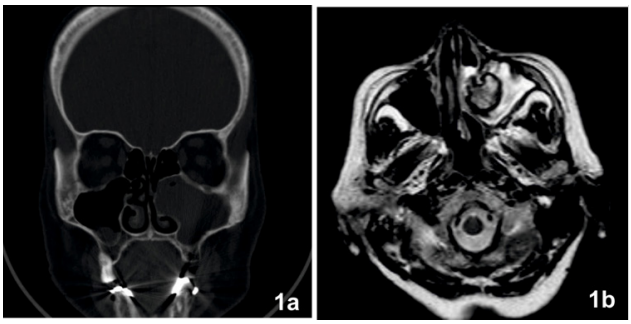

Figure 1. (a) Coronal CT scan of paranasal sinüs showing the mass in the left maxillary sinus cavity, causing a scalloping on the medial wall of the sinus and narrowing the left osteomeatal unit and nasal cavity. (b) Axial MRI show tumoral mass lesion filling the left maxillary sinüs.

amyloidosis and localized amyloidosis. Primary systemic amyloidosis is a systemic condition with no known underlying reason. Secondary systemic amyloidosis occurs with underlying medical problems such as multiple myeloma, tuberculosis and rheumatoid arthritis. Renal and cardiac diseases, which are the most frequent causes of death, are seen in both the primary and secondary forms (3). The 3rd form of amyloidosis is the localized form and there is no underlying disease or systemic involvement. This form is very rare compared to the other forms (2).

Amyloid depositions in the head and neck region can be a part of localized or systemic amyloidosis (4). Localized amyloidosis in the head and neck region is a rare phenomenon and any organ in the head and neck region can be affected. Larynx is the most frequently involved site in the head and neck region, followed by oropharynx, trache, orbit, sinonasal tract and nasopharynx (2). Isolated sinonasal amyloidosis is an extremely rare condition and has been reported in a limited number of patients (4). Having a localized amyloidosis involving the maxillary sinus, our case represents a very rare condition too. When the literature is reviewed, the number of cases having localized sinonasal amyloidosis is only 15 until 2012 (5).

Symptoms of head and neck amyloidosis vary based on the region affected. The most common head and neck presentations are hoarseness, nasal congestion, odynophagia, articulation problems, mandibular deformities, deglutition difficulties, airway obstruction, speech disorders and hypogeusia (3). Sinonasal amyloidosis presents with nasal obstruction, nasal discharge, epistaxis (due to vessel wall invasion), middle ear effusion and post nasal drip (2). Our case is came with left-sided nasal obstruction and intermittent epistaxis. The appearance is a yellowish or whitish polypoid, firm mass. It can be easily confirmed by tissue biopsy (2). Eosinophilic extracellular deposits of protein fibrils staining with Congo red and exhibiting apple-green birefringence when viewed under polarized light is the mainstay of the diagnosis (6). After establishing the diagnosis, a detailed examination is required for systemic amyloidosis. This examination should include biopsy of the rectum and abdominal fat biopsy. These tests are positive in 75-90% of the patients with systemic amyloidosis (7). Serum or urine Bence-Jones proteins are found in up to 88% of patients with primary systemic amyloidosis and 100% of patients with multiple myeloma-associated systemic amyloidosis (8). An echocardiogram can be obtained to evaluate the myocardium for signs of amyloidosis. Moreover, additional tests such as dynamic magnetic resonance imaging, Tc-99m phosphate radionuclide imaging and 123I serum amyloid P scintography can be useful in evaluating the presence and spread of the disease (9). Our patient was diagnosed as localized maxillary sinus amyloidosis upon performing a biopsy with total excision of the mass, observing a characteristic appearance with Congo staining and excluding systemic involvement upon a detailed examination.

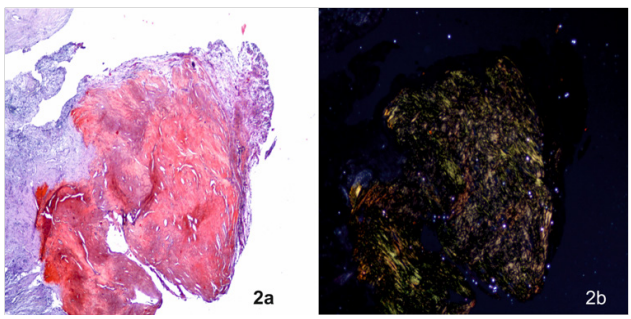

Figure 2. (a) Congo red stain of amyloid (40x magnification). (b) Characteristic green birefringence with polarized light (40x magnification).

Surgical excision is the treatment for localized amyloidosis. Repeated operation may be indicated for residual or multifocal disease. Bleeding can be seen as a complication in some cases. A close follow-up is required as the recurrence rate of the disease is high (2). Radiotherapy, chemotherapy and corticosteroids are regarded as ineffective (4). In our patient, the mass excision was performed by endoscopic medial maxillectomy. No post-operative complication was encountered and no recurrence was observed after the 6 months follow-up period. Localized amyloidoma of the maxillary sinus is a very rare condition. Systemic involvement should be excluded through a detailed examination as it affects survival in such patients.

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