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A CASE OF MIGRATORY ORBITAL INFLAMMATION: KIMURA'S DISEASE

Case Report

ORBİTAL MİGRATUAR İNFLAMASYON İLE SEYREDEN BİR OLGU: KİMURA HASTALIĞI

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ÖZET

Amaç

Kimura hastalığı, etyolojisi bilinmeyen, nadir görülen, benign natürde ve kronik bir hastalıktır. Erkeklerde 6 kat daha sık rastalanan, ilk önceleri de "eozinofilik hiperplastik lenfogranüloma" şeklinde isimlendirilmiş bu patolojide; baş-boyun bölgesinde, ciltaltı dokuda kitlesel olmayan inflamatuar kalınlaşmalar meydana gelmektedir. Biz bu sunumda, orbital tutulumu bulunan bir olgunun görüntüleme bulgularını paylaşmayı amaçladık.

Materyal Metod

Sol göz üst kapak altı şişlik ve hafif ağrı ile başvuran olguda kontrastlı orbita MRG incelemesi yapılmış, daha sonra biopsi alınmış ve 7 ay sonra da sağ göz arkasında aynı şişlik ve ağrı benzeri tablo ile başvuran olguda (49 yaş, kadın olgu) orbita MRG inceleme yapılmış ve değerlendirmeler bu görüntüler üzerinden gerçekleştirilmiştir.

Bulgular

Hastanın ka beyaz küre değerleri ılımlı derecede yüksek olup, literatür bilgilerinin aksine, çok belirgin bir eozinofil artışı tespit edilmemişti. Ancak literatür ile uyumlu olarak; olgunun fiziki muayene bulgularında da hafif derecede ağrı yakınması dışında herhangi bir bulgu tespit edilmemişti. Diğer taraftan ilk MRG görüntülerinde sol anterosüperior preseptal alanda, kontrol MRG'sinde ise ilaveten sağ retroorbital mesafede; yağlı planlarda inflamasyon ve kontrast tutulumu tespit edilmişti. Kesitsel inceleme orbital psödotümör ile uyumlu iken, yaygın inflamatuar yağlı doku ile çevrelenmiş ekstraoküler kas grupları hiç etkilenmemiş, normal kalibrasyonda, normal sinyal özelliklerinde idi. Olgunun ilk başvurusunda alınmış patolojisi Kimura Hastalığı ile uyumlu olarak rapor edilmişti.

Sonuç

Bu sunum ile, atipik klinik ve radyolojik karakterler sergileyen Kimura Hastalığı tanısı almış bir olguda, nadir görülmesine karşın bu hastalıktan şüphelendirebilecek özellikleri ortaya koyduk.

Anahtar Kelimeler: *Kimura; Lenfogranülom; Lenfadenopati; orbita inflamasyonu.*

SUMMARY

Purpose

Kimura's disease is a rare, benign and chronic disease of unknown etiology. This pathology has 6-fold higher prevalence in males and has been initially referred to as "eosinophilic hyperplastic lymphogranuloma"; it is manifested by inflammatory thickening of subcutaneous tissue in the head and neck. In this case report, we aimed to share imaging findings of a patient with orbital involvement.

Materıal and Method

The patient presented with swelling of the left upper eyelid and mild pain; the patient underwent orbital MRI with contrast enhancement and biopsy was performed. She (49-year-old, female) was re-admitted 7 months later due to similar swelling in the right eye and pain; and then underwent orbital MRI and assessments were carried out on these images.

Results

The patient's white blood cell count was moderately high and, in contrast to the literature, eosinophil count was not markedly elevated. However, physical examination did not reveal any abnormality excluding mild pain which is in line with the literature. On the other hand, initial and control MR images have revealed inflammation of fat tissue and contrast uptake in the left anterosuperior preseptal area and in the right retroortibal space, respectively. Cross-sectional studies were consistent with orbital pseudotumor whereas extraocular muscle groups surrounded by extensive inflammatory fatty tissue were not affected and they were of normal calibration and signal intensity. Pathologic examination performed after the first admission of the patient was consistent with Kimura's disease by the appearance of vascular proliferation with eosinophylic infiltration.

Conclusion

In this case report of a patient who was diagnosed with Kimura's disease that represents atypical clinical and radiological characteristics, we present rare features which, however, may raise the suspicion of this disease.

Keywords: *Kimura; Lymphogranulom; Lymphadenopathy;orbital inflammation.*

INTRODUCTION

Kimura's disease is a rare, benign and chronic inflammatory disease of unknown etiology. This pathology has 6-fold higher prevalence in males and has been initially referred to as "eosinophilic hyperplastic lymphogranuloma"; it is manifested by inflammatory thickening of subcutaneous tissue in the head and neck (1). Peripheral eosinophilia, elevated serum levels of IgE and swelling in the head and neck form the characteristic triad of the disease (1, 2). Clinically, the disease is characterized by lymphadenopathy and painless subcutaneous nodules or masses which sometimes cause widespread itching that result in skin erosions. The lesions may show recurrence or disappear spontaneously. Some lesions may recur after months or even years. In this case report, we aimed to share imaging findings of a patient with orbital involvement.

MATERIAL AND METHOD

The patient (49 year-old female) presented with the complaints of swelling beneath the left upper eyelid and mild pain; the patient underwent orbital MRI with contrast enhancement and biopsy was performed. She was re-admitted 7 months later due to similar swelling in the right eye and pain and then underwent second orbital MRI using gadolinum-based contrast agent. Post-contrast sequences with fat suppression were obtained along with conventional pre-contrast and fat-suppressed T1-T2-weighted conventional multiplanar sequences.

RESULTS

The patient's white blood cell count was moderately high (13.200/mm3) and blood eosinophil levels were below the upper limit of normal ranges. In line with the literature, physical examination of the patient showed mild-to-moderate pain. On the other hand, initial and control MR images had revealed inflammation of fat tissue and contrast uptake in the left anterosuperior preseptal area and in the right retroortibal space, respectively **(Figure 1).**

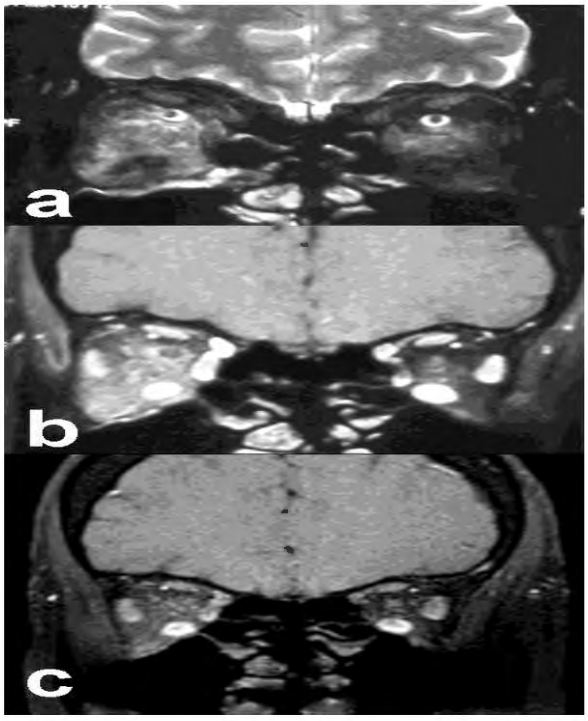

Figure 1. *a) Coronal fat-suppressed T2-weighted sequence, b) Increased signal intensity and contrast uptake are observed in coronal fat-suppressed T1-weighted contrast-enhanced sequences of bilateral retroorbital fat tissue that is more prominent on the left side. c) Posterior sequences did not show marked involvement of extraocular muscle groups or optic nerve except minimal reactive contrast uptake.*

These changes had non-mass character. Despite the presence of inflammation and infiltration, there was no obvious evidence of invasion. Cross-sectional studies were consistent with orbital pseudotumor whereas extraocular muscle groups surrounded by extensive inflammatory fatty tissue were not affected and they were of normal calibration and signal intensity. Histopathologic examination of the surgical biopsy samples obtained on initial admission presented as eosinophilic infiltration and vascular proliferation with fibrosis and was reported to be consistent with Kimura's disease.

DISCUSSION

The pathophysiology of Kimura's disease remains unknown, although an allergic reaction, trauma, and an autoimmune process have all been implicated as possible causes. It is a chronic inflammatory condition involving skin and subcutaneous tissue of the head and neck and rarely oral mucosa and mostly affects young-middle aged men. Histopathologically, Kimura's disease manifests lymphocytic infiltration and eosinophilic infiltration of lymphoid follicles, vascular proliferation (proliferation of postcapillary venules) and fibrosis (2, 3). The most common histologic features of Kimura disease include preserved nodal architecture; follicular hyperplasia with reactive germinal centers; well-formed mantle zones; eosinophilic infiltrates involving the interfollicular areas, sinusoidal areas, perinodal soft tissue, and subcutaneous tissue; and proliferation of postcapillary venules(2, 3). Detailed laboratory data of our patient was not accessible (due to

residence in a foreign country). Therefore, blood IgE levels were not known. However, histopathological data had been reported to be consistent with Kimura's disease.

Involvement of other organs (kidney, lungs, etc.) has not been found in Kimura's disease. However, it must be realized that this disease may also show involvement of other organs (4). Kimura's disease is most often confused with angiolymphoid hyperplasia. Angiolymphoid hyperplasia is characterized by angiomatous papules and nodules localized especially in head and neck and presents in middle aged women with peripheral eosinophilia and elevated serum IgE levels. The differential diagnosis of the disease in these two similar diseases is only possible with histopathologic examination (5). As is the case in our patient, in circumstances where there is atypical orbital inflammatory involvement, it must be kept in mind that radiological features of Kimura's disease could be confused with other inflammatory-infectious conditions or with malignant neoplasms.

CONCLUSION

In this case report, we attempted to describe involvement of orbital plains together with the findings on MRI with contrast enhancement in a patient who was diagnosed with Kimura's disease that represents atypical clinical and radiological features.

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