Cukurova Medical Journal
ÇUKUROVA ÜNİVERSİTESİ TIP FAKÜLTESİ

Cukurova Med J 2022;47(2):901-904
DOI: 10.17826/cumj.1063305

EDİTÖRE MEKTUP / LETTER TO THE EDITOR

A rare case of Mönckeberg medial calcific sclerosis simulating giant cell arteritis

Dev hücreli arteriti taklit eden nadir bir Mönckeberg medial kalsifik skleroz olgusu

Burak Okyar[1], Bekir Torun[1], Fatih Albayrak[1], Sezen Koçarslan[2], Fatih Yıldız[1], Gözde Yıldırım Çetin[1]

[1]Kahramanmaraş Sütçü İmam University, Departmant of Rheumatology, [2]Departmant of Pathology, Kahramanmaraş, Turkey

Cukurova Medical Journal 2022;47(2):901-904

To the Editor,

Giant cell arteritis (GCA) is a chronic, idiopathic, granulomatous, and inflammatory vasculitis involving medium to large vessels. It mainly occurs over 50 years of age and is often affected by the temporal artery and its branches[1]. Clinical findings of temporal artery biopsy and temporal artery doppler ultrasonography (d-USG) in the diagnosis of GCA are still the gold standard[2]. In the biopsy, necrosis foci, intimal hyperplasia, epithelioid cell, multicore giant cell, internal elastic lamina destruction are observed in the vascular wall. It is characterized by granulomatous inflammation, tunica media, and tunica intima, holding all the layers and narrowing the lumen. In d-USG, narrowness, obstruction, and hypoechoic halo (halo sign) around the artery and especially intimal wall thickening leads to the diagnosis of inflammatory arthritis[3].

Johan Georg Mönckeberg defined Mönckeberg Medial Calcific Sclerosis (MMCS) in 1903 as calcification that occurs with the accumulation of hydroxyapatite crystals in the tunica media layer of small and medium-sized vessels and the elastic internal/external lamina[4]. Although many pathophysiological mechanisms have been suggested, its etiology has not been fully elucidated. MMCS is a subtype of vascular calcification and is characterized by calcification of the tunica media layer. This calcification focus is anatomically separated from the calcification that develops in the atheroma plate, which is the other type of vascular calcification, and involves the tunica intima layer[5]. However, in severe MMCS patients, the tunica intima layer may also be calcified. Sometimes, calcification is severe, and calcification develops in the internal/external elastic lamina, destroying the elastic lamina. In the pathological interpretation of the biopsy material, this may mislead the clinician, assuming it is the sequelae of previous inflammatory arthritis[6]. MMCS, which is mostly asymptomatic clinically, is detected incidentally on radiographic imaging. Train track appearance is the most common finding in plain radiographs[7]. In cases diagnosed and reported as MMCS, the onset is most common in the arteries of the lower extremities. It then involves the upper extremity arteries and the visceral arteries[8]. In severe cases, it causes ischemia, especially in the lower extremities, causing non-healing ulcerated wounds and amputations. It occurs most frequently in patients with type 2 diabetes mellitus (DM) (17.8%)[11]. In addition, advanced age, male gender, end-stage renal disease (ESRD), dialysis patients, osteoporosis, vitamin K antagonist use, hyperparathyroidism are predisposing factors. Rarely, it may develop secondary to rheumatoid arthritis and Kawasaki disease[12].

In this case, we present a rare case that mimics GCA but was diagnosed as MMCS with temporal artery

Yazışma Adresi/Address for Correspondence: Dr. Burak Okyar, Kahramanmaraş Sütçü İmam University, Department of Rheumatology, Kahramanmaraş, Turkey E-mail: okyarmd@gmail.com
Geliş tarihi/Received: 26.01.2022 Kabul tarihi/Accepted: 27.02.2022

involvement with clinical, radiological, and pathological findings.

A 56-year-old female patient presented with sudden-onset right vision loss. In the history, there was a severe headache in the region corresponding to the temporal artery trace on the same side and scalp sensitivity in the same region. The patient had jaw pain caused by chewing. The patient had a history of hemodialysis (HD) and hypertension for 13 years. Both big toes had been amputated due to previous ulcerated wounds. In physical examination; There was significant scoliosis and tenderness in the temporal artery trace. Peripheral pulses could not be obtained from all four extremities of the patient. In the ophthalmic examination, the right optic disc was pale and bilateral maculae were normal. Right-sided vision was 63%, left-sided 96%. In routine blood tests; White Blood Cell: 7.86x109/L, Hemoglobulin: 10.9 g/dL, platelet: 410x103 U/L, C-reactive protein: 3.03 mg/L, Sedimentation: 53 mm/hour, Creatinine: 3.16 mg/dL, Calcium: 9.95 mg/dL, Phosphorus: 2.85 mg/dL, Glucose: 96 mg/dL. Other tests of the patient were within normal limits. Bilateral temporal artery d-USG; There was irregular wall thickening, more prominent on the left side.

Arterial lumens were clear. No additional pathology was detected in the patient in orbital magnetic resonance imaging (MRI) and cranial MRI examinations. A temporal artery biopsy was performed on the patient with a preliminary diagnosis of GCA. A 3 cm long artery biopsy was evaluated: diffuse foci of calcification were detected in the tunica intima and tunica media layers, which distorted the vascular smooth muscle layer. There was mild neutrophilic infiltration in the tunica adventitia layer (Figure-1). Existing histomorphological findings were reported as arteritis developing based on chronic atherosclerosis. Considering giant cell arteritis, 48 mg methylprednisolone treatment was started in the patient, and he was followed up.

In the dual X-ray bone densitometry performed in the patient with diffuse bone pain in the control examinations, a T-score of -4.8 was detected at the L1-L4 level (Figure-3). On the radiographs of four extremities, calcified arterial appearance, giving the appearance of a train rail, was detected in the patient (Figure-2,3). Parathyroid hormone level was measured as 5805 ng/L. He had ulcerated sores on his feet that did not heal. In lower extremity arterial doppler ultrasonography, Pulsation was not observed in bilateral popliteal and distal arteries, but the flow was detected. Diffuse calcifications were detected in the arterial structure in the bilateral lower extremity. Although the patient had a giant cell arteritis clinic when all the findings were combined, MMCS was diagnosed when the patient's comorbid diseases, pathological findings, radiological images, and clinic were evaluated together.

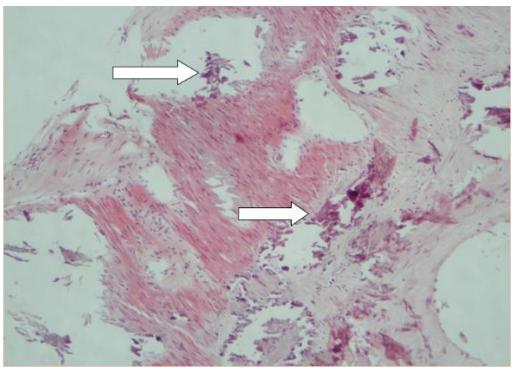

Figure 1. Calcification areas distorting and dissecting the muscle structures are observed in the media layer in the section of the artery wall, the lumen of which cannot be seen due to obstruction (H&E X100 objective).

In this case report, a patient who presented with a clinical diagnosis of GCA was diagnosed with MMCS, a rare pathology in the literature. This case is the 5th known case with GCA clinic and diagnosed as MMCS in the literature. The clinician should well evaluate clinical, radiological, and pathological findings to diagnose MMCS.

In 131 temporal artery biopsy samples between 1975-1998 by Illinois University Chicago eye center; Atherosclerosis with myointimal fibrosis was diagnosed in 63%, giant cell arteritis in 13%, and calcific sclerosis in 6%[13]. This study shows that some cases diagnosed with GCA are MMCS. A literature review shows that MMCS is more common in patients with diseases such as type 2 DM, ESRD, hyperparathyroidism, and osteoporosis[14]. In our case, all of these pathologies were present except type 2 DM. MMCS starts mainly in the lower extremity and progressive causes; the visceral arteries and the upper extremity are involved, and rarely the coronary, thyroid, and temporal artery regions[14]. This information indicates that MMCS is a systemic pathology. In our case, the lower extremity, upper extremity, digital arteries of the hands, and temporal artery were also affected (Figure 2,3). Biopsy findings for GCA are the standard gold method. Giant cells,

epithelioid histiocytes, granulomas, and fragmented elastic lamina are findings leading to the diagnosis[3]. None of these were detected in our case, and tunica media calcification and degradation of vascular smooth muscle led to the diagnosis of MMCS.

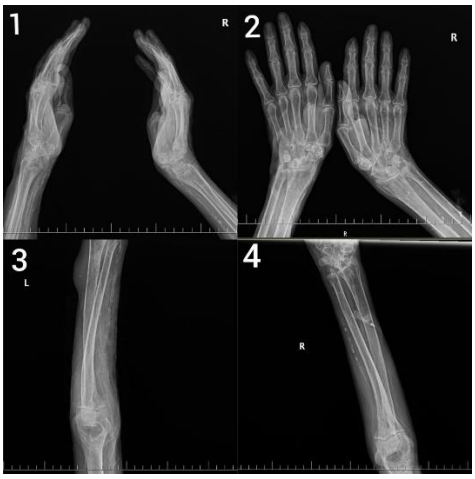

Figure 2. Arterial calcification in both upper extremities giving train-rail appearance. Digital, radial and ulnar artery calcifications can be clearly distinguished.

The sudden loss of vision that developed in the patient was explained by the diagnosis of non-arteritic ischemic optic neuropathy in the ophthalmological examination performed during the patient's follow-up. In the control fundus examination, optic atrophy was detected on the patient's right side in the progressive process, and pallor was detected in the optic disc at the left base of the eye. This finding shows that MMCS is a pathology that is serious enough to cause vision loss with ischemia that develops after temporal artery involvement and shows that it can completely mimic GCA. Francisco et al. published the results of 4 MMCS cases in 2020. All 4 cases diagnosed with MMCS had visual impairment, permanent visual impairment was observed in 2 cases, and temporary impairment was observed in 1 case[1]. Although this 4-case review shows that the results of MMCS can be permanent and severe, permanent vision loss developed in our case as well.

Conventional radiographs are essential for suspecting MMCS. Train track appearance, especially in the lower extremities, is one of the first findings to make us suspect MMCS[8]. Only upper extremity involvement without lower extremity involvement is infrequent in the literature. Therefore, lower extremity involvement is a finding that will strengthen our diagnosis. In our case, both lower extremities were affected, and the train-rail appearance supported our diagnosis. MMCS may cause loss of elasticity of the arteries, resulting in loss of pulse and extremity ischemia. In our case, arterial pulsation loss was detected by physical examination and d-USG. Non-healing ulcerated wounds occurred in both feet as a result of limb ischemia. MMCS is characterized by calcification of the tunica media layer[1]. Although inflammatory cell infiltration is not expected, an autopsy study on Tamandua Tetradactyla in a zoo in 2021 revealed tunica media calcification and very mild neutrophil infiltration in the tunica intima layer[15]. This finding shows that inflammatory cell migration to the arterial wall may occur in the pathological process of MMCS and may lead clinicians to diagnose inflammatory artheritis. However, in our case, there was minimal neutrophilic infiltration in the tunica adventisia layer.

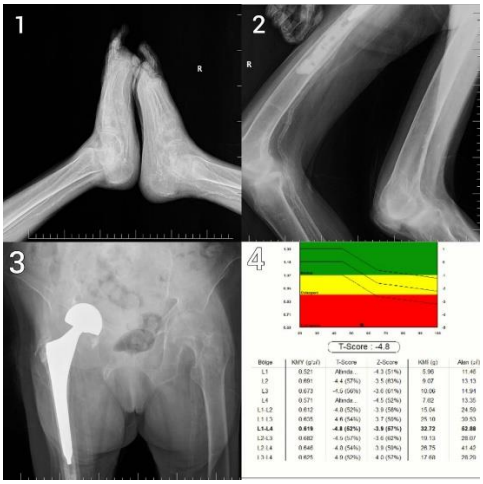

Figure 3. Train track view is seen in the arteries of both lower extremities. Calcifications can be detected in the trace extending to both external iliac arteries, popliteal arteries, tibialis posterior, dorsalis pedis and metatarsal arteries. The patient's bone densitometry measurement is seen in the image number 4.

In conclusion, with this case report, a patient presented with the GCA clinic was diagnosed with MMCS, a vasculitis mimic. Knowing the radiological, clinical, and pathological manifestations of MMCS is of great importance in guiding clinicians correctly.

Yazar Katkıları: Çalışma konsepti/Tasarımı: GYÇ, FY; Veri toplama: BO; Veri analizi ve yorumlama: BT; Yazı taslağı: BO; İçeriğin eleştirel incelenmesi: GYÇ, FY; Son onay ve sorumluluk: BO, BT, FA, SK, FY, GYÇ; Teknik ve malzeme desteği: SK; Süpervizyon: GYÇ; Fon sağlama (mevcut ise): yok.
Etik Onay: Bu çalışma editöre mektup niteliğinde olup, etik kurul onayına gerek yoktur. Hasta aydınatılmış onam vermiştir.
Hakem Değerlendirmesi: Editoryal değerlendirme.
Çıkar Çatışması: Yazarlar çıkar çatışması beyan etmemişlerdir.
Finansal Destek: Yazarlar finansal destek beyan etmemişlerdir.

Author Contributions: Concept/Design : GYÇ, FY; Data acquisition: BO; Data analysis and interpretation: BT; Drafting manuscript: BO; Critical revision of manuscript: GYÇ, FY; Final approval and accountability: : BO, BT, FA, SK, FY, GYÇ; Technical or material support: SK; Supervision: GYÇ; Securing funding (if available): n/a.
Ethical Approval: This study is a letter to the editor, and ethics committee approval is not required. Patient provided informed consent.
Peer-review: Editorial review
Conflict of Interest: Authors declared no conflict of interest.
Financial Disclosure: Authors declared no financial support

REFERENCES

1. Cuevas Castillo FJ, Sujanani S, Chetram VK, Elfishawi M, Abrudescu A. Monckeberg Medial Calcific Sclerosis of the temporal artery masquerading as giant cell arteritis: case reports and literature review. Cureus. 2020;12:e9210.
2. Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum. 1990;33:1122-8.
3. Schmidt WA, Kraft HE, Vorpahl K, Völker L, Gromnica-Ihle EJ. Color duplex ultrasonography in the diagnosis of temporal arteritis. N Engl J Med. 1997, 337:1336-1342.
4. Lanzer P, Boehm M, Sorribas V, Thiriet M, Janzen J, Zeller T et al.: Medial vascular calcification revisited: review and perspectives. Eur Heart J. 2014, 35:1515-25.
5. Lachman AS, Spray TL, Kerwin DM, Shugoll GI, Roberts WC. Medial calcinosis of Mönckeberg. A review of the prob- lem and a description of a patient with involvement of periph- real, visceral and coronary arteries. Am J Med. 1977;63:615-22.
6. Meema HE, Oreopoulos DG, Rapoport A. Serum magnesium level and arterial calcium cation in end-stage renal disease. Kidney Int. 1987;32:388-94.
7. Silbert S, Lippmann HI: Mönckeberg's arterioclerosis. JAMA. 1955;151:1176-79.
8. Castling B, Bhatia S, Ahsan F. Mönckeberg's arteriosclerosis: vascular calcification complicating microvascular surgery. Int J Oral Maxillofac Surg. 2015;44:34-6.
9. Lehto S, Niskanen L, Suhonen M, Ro ¨nnemaa T, Laakso M. Medial artery calcification: a neglected harbinger of cardiovascular complications in non-insulin-dependent diabetes mellitus. Arterioscler Thromb Vasc Biol. 1996;16:978 –093.
10. Couri CE, da Silva GA, Martinez JA, Pereira Fde A, de Paula FJ. Mönckeberg's sclerosis - is the artery the only target of calcification? BMC Cardiovasc Disord. 2005;5:34.
11. Castillo BV Jr, Torczynski E, Edward DP. Monckeberg's sclerosis in temporal artery biopsy specimens. Br J Ophthalmol. 1999;83:1091-2.
12. Lanzer P, Boehm M, Sorribas V, Thiriet M, Janzen J, Zeller T et al. Medial vascular calcification revisited: review and perspectives. Eur Heart J. 2014;35:1515-25..
13. Kimura T, Inaka K. Arterial medial calcification (Mönckeberg's sclerosis) with chronic renal disease in a zoo-kept Southern tamandua (Tamandua tetradactyla). Int J Vet Sci Med. 2021;9:1-6.