Case Report

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Pathology Section

Monckeberg's Calcification as a Clinical Mimicker of Temporal Arteritis: A Case Report

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ABSTRACT

Temporal arteritis usually presents in patients above 50 years and is characterised by granulomatous inflammation in all layers of the arterial wall. On the other hand, Monckeberg medial calcification is a disease condition limited to the tunica media of the affected vessels. In this report, authors present the case of a 68-year-old man who presented to the casualty with facial pain. He also had a runny nose, cough, and sleep disturbance. A Computed Tomography (CT) scan was performed and reported as normal. The patient underwent Ear, Nose and Throat (ENT) and Ophthalmology evaluations. Neurology opinion revealed tenderness and thickening of the right superficial temporal artery. Based on clinical suspicion, the patient was started on oral corticosteroids, suspecting temporal arteritis. A temporal artery biopsy was performed for histopathological examination, which surprisingly revealed no evidence of arteritis but only calcification of the tunica media. Unfortunately, the patient was lost to follow-up after the biopsy. This case highlights the importance of recognising Monckeberg medial calcification in clinically suspected cases of temporal arteritis, as there are only a few reported cases in the literature.

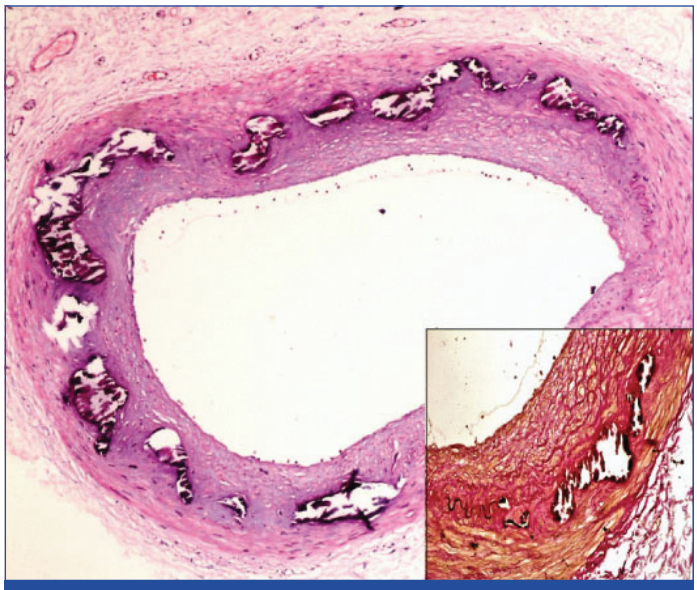
Keywords: Monckeberg medial calcification, Persistent headache, Temporal artery biopsy

CASE REPORT

A 68-year-old male patient came to the casualty with severe continuous craniofacial pain on the right-side for five days, along with sleep disturbance. He also had a runny nose and cough for the same duration. The patient had a medical history of chronic obstructive pulmonary disease, hypertension, chronic kidney disease, and was recently diagnosed with Type 2 Diabetes during his hospital stay, and he started taking medications for it. He has been a smoker for the past 20 years. A past history of trauma to the right-side of his head five years ago was elicited, but the CT scan performed at that time showed no abnormalities. During the examination, diffuse tenderness was found on the right-side of his face, specifically over the right temporal artery region. No specific tenderness was noted over the paranasal sinuses or orbit. An ENT evaluation was obtained, and the opinion given was that it was resolving acute sinusitis. However, since the patient had persistent headaches, an opinion from an ophthalmologist was sought, and the examination revealed no disc oedema. In view of the persistent headache, a neurologist's opinion was also sought, who identified tenderness and thickening of the right superficial temporal artery, indicating the possibility of temporal arteritis. Oral corticosteroid, prednisolone, was started at a dose of 60 mg/day, taking into consideration his weight. An Magnetic Resonance Imaging (MRI) examination of the brain revealed diffuse age-related cerebral atrophy, small vessel ischaemic changes, and bilateral sinusitis.

A right temporal artery biopsy was performed, and the pathologist received a segment of the vessel measuring 0.5 cm in length. The vessel diameter and lumen diameter measured 0.2 cm and 0.1 cm, respectively. Microscopic examination [Table/Fig-1] showed extensive Monckeberg medial calcification in the vessel. Inflammatory cells were not detected in the vessel wall. Elastic stain revealed the absence of the internal elastic lamina in the majority of zones, with fragmentation observed in the remaining areas.

Oral corticosteroid treatment with a dose of 60 mg/day of prednisolone was initiated considering the diagnosis of temporal arteritis at the time of discharge. However, the patient was lost to follow-up, and this treatment did not prove to be beneficial as per the histopathological examination that revealed Monckeberg's calcification.



[Table/Fig-1]: Extensive Monckeberg calcification in the vessel wall with no evidence of inflammation (Haematoxylin & Eosin, 100x). In Inset box- Elastic stain showed absence of internal elastic lamina in majority of zones along with fragmentation in the remaining areas; (Verhoeff- Van Gieson stain, 400x).

DISCUSSION

Temporal arteritis is a rare disease with an incidence of 9.3 per 100,000 population and a histologically proven incidence of 5.5 per 100,000 population [1]. The clinical features are vague and constitutional, such as fever, fatigue, and weight loss, along with facial pain/headache. The pain is most intense along the course of the superficial temporal artery, which is tender on palpation [2]. Pathologically, the changes in affected vessels are patchy along their length. The majority of cases show granulomatous inflammation and fragmentation of the internal elastic lamina, while a minority exhibit non specific panarteritis [2]. In the present case, the patient was strongly suspected to have temporal arteritis based on clinical grounds. Temporal artery biopsy, which remains the gold standard

for diagnosis, was performed [3]. However, microscopic examination revealed an entirely different finding: extensive Monckeberg medial calcification with no features suggestive of temporal arteritis. Monckeberg arteriosclerosis of the temporal artery can occasionally be observed.

To the best of our knowledge, only a few cases of Monckeberg's calcification in patients with clinically suspected temporal arteritis have been reported. Three articles have documented four cases based on temporal artery biopsies. Three of these cases involved individuals above 50 years of age who complained of headaches, with two of them experiencing tenderness in the temporal area. One patient presented with acute vision loss and temporal artery hardening [4-6].

In a study involving 131 pathology specimens of temporal artery biopsies, it was found that 63% exhibited atherosclerosis with intimal fibrosis, 13% had giant cell arteritis, and 6% showed Monckeberg calcification [7]. Some authors have observed an association between Monckeberg's calcification and temporal arteritis in patients with diabetes mellitus [8] and chronic kidney disease [8]. Previous trauma has also been noted as a potential association. Index patient had a history of trauma and diabetes mellitus. This histological diagnosis is interesting but has little clinical significance, although it should be recognised in temporal artery biopsy specimens. The exact relationship between Monckeberg calcification and clinical findings is unknown, and it responds poorly to oral corticosteroids. Therefore, it is suggested that biopsy should be performed in all cases of clinically suspected temporal arteritis to determine the true incidence of the association between Monckeberg calcification and the course of the disease.

CONCLUSION(S)

The presence of Monckeberg's calcification in the temporal artery biopsy of a patient clinically suspected of temporal arteritis is an intriguing histopathological diagnosis. Treating physicians and pathologists should be aware of this condition in temporal artery biopsies. Identifying this condition alters the management of cases with suspected temporal arteritis. Further studies are also required to determine the association of this condition with diabetes mellitus. Identification of this condition changes the management of the cases with suspected temporal arteritis. Further studies are also required to know the association of this condition with diabetes mellitus.

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