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ATRIAL MYXOMA - A RARE CAUSE OF STROKE (CASE SERIES)

Neurology

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ABSTRACT

Most strokes in young adults are ischemic. Atrial myxoma's are a rare cause of stroke, accounting for fewer than 1% of all ischemic strokes. Myxomas are commonest cardiac tumors mostly seen in left atrium. However, left atrial myxoma presenting with stroke is rare clinical condition whose etiology is unknown. Neurological sequelae after resection are rare but may occur without recurrence of the cardiac tumor. Detection of the tumor by echocardiography, followed by surgical excision, significantly reduces the risk of subsequent ischemic strokes.

KEYWORDS

Atrial Myxoma, Stroke, Left Atrium, Infarct

Introduction:

Stroke is generally considered to be a disease of middle-aged and older adults however it can occur in young adults as well. Data collected in Europe and the United States have demonstrated an annual incidence of 4 to 28 stroke events per 100,000 people younger than 45 years [1]. Most strokes in young adults are ischemic. Atrial myxoma's are a rare cause of stroke, accounting for fewer than 1% of all ischemic strokes [2]. Detection of this tumor is relatively easy, and surgical removal of the myxoma is usually a permanent measure to prevent subsequent strokes. Cardiac myxoma is a source of emboli to the central nervous system and elsewhere in the vascular tree. However, nonspecific systemic symptoms and minor embolic phenomena may be overlooked in the absence of any history of cardiac problems. In this situation, cardiac investigations may not be performed, and diagnosis of this rare condition may be delayed until the onset of more significant embolic disease, such as stroke with functional impairment, as in the case reported here. Myxomas are a rare cause of ischemic stroke accounting for less than 1% of all stroke cases. However, they should be considered in the differential diagnosis of cases with stroke in young in the absence of conventional stroke risk factors [3].

Case Presentation 1:

A 43-year-old woman with no co-morbid illness was admitted with history sudden onset of right sided weakness with speech difficulty for 1 day. There was no history of headache, fever, seizures, or trauma to head. She had history of similar weakness lasting for few minutes to hours for four times over last few months. Her neurological examination revealed motor aphasia, right UMN facial nerve palsy and hemiparesis of right side of the body. Right plantar reflex was extensor. Rest of nervous system and other systemic examination was unremarkable except for a mid-diastolic mitral murmur on cardiac auscultation and it was heard with the postural variation.

Her blood picture and biochemical parameters reports were normal. ECG showed non-specific ST-T changes. MRI showed a large acute non-hemorrhagic infarct in the left temporo-parietal region in the left middle cerebral artery territory causing mass effect on left lateral ventricle. Chest X ray suggest left atrial enlargement. In view of stroke in the young with cardiac mid-diastolic murmur and chest x ray findings, echocardiogram was done which showed enlarged left atrium with an echogenic pedunculated mass (6.1x3.2 cm) attached to septum, highly suggestive of Atrial Myxoma (Figure -1).

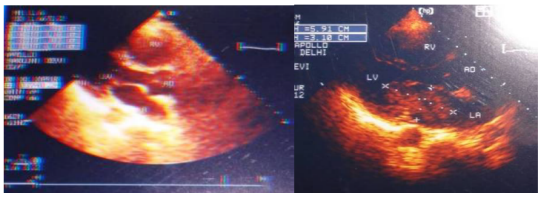

Figure : 2D Echo showed enlarged left atrium with an echogenic pedunculated mass (6.1x3.2 cm) attached to septum (Atrial Myxoma).

Case Presentation 2:

A 39 years old man presented to ER with sudden onset loss of consciousness, right upper limb and lower limb weakness with inability to speak since 2 days. There was no history of fever, headache or trauma. No history of similar complaints in the past. His neurological examination revealed motor aphasia and right hemiparesis. Right plantar was extensor. Cardiac auscultation revealed third heart sound with tumor plop murmur, and it was heard with the postural variation. Rest nervous system and other systemic examination was unremarkable.

His routine blood tests, ECG and Chest X- ray were fairly normal, with no significant abnormalities. MRI brain was done which revealed acute ischemic infarct in the left middle cerebral artery territory (Figure 2). In view of the stroke in the young with cardiac murmur, a complete cardiac work up was done. 2D echocardiography revealed enlarged left atrium with an echogenic mass (5.2 x 2.7 cm), which was highly suggestive of atrial myxoma tumor.

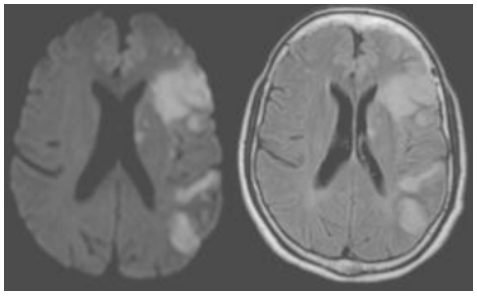

Figure 2 - acute ischemic infarct in the left middle cerebral artery territory

Treatment:

Both the patients were eavlauated for atrial myxoma. Patients were attached to cardiothoracic surgery for surgical treatment. There was initial controversy in operating the left atrial myxoma as an emergency. Review of literature revealed that there are no strict guidelines regarding the management of acute stroke in case of left atrial myxoma. However, those few cases which have been dealt with at centre's worldwide advised that it is better not to intervene in first 48 hours after stroke. It is safer to manage the acute cerebral infarct. In view of this, we managed these patients with standard stroke care treatment and anti-edema measures like mannitol for a couple of days with a repeated CT scan to assess the cerebral infarct zone. First patient

(Case 1) underwent cardiac surgery for removal of atrial myxoma after one week of treatment. Second patient (Case 2) underwent similar surgery after 6 days of admission for removal of atrial myxoma. The histopathological examination in both the cases showed features consistent with diagnosis of atrial myxoma. Both the patients recovered over the period of next few weeks except for residual right sided hemiparesis and were discharged in a stable condition. Both patients are free of stroke recurrence in the follow up of post-operative period of more than a year.

Discussion:

Primary cardiac tumors are extremely rare and constitute only about 5% of all cardiac tumors [4–7]. Cardiac myxomas are noncancerous primary tumors of the heart and constitute about of 50% of all primary heart tumors [7]. Approximately 75% of them arise from the left atrium, 20% of them from the right atrium, and 5% in both atria and the ventricle [5]. Cardiac myxomas predominantly occur in women [8] with an average age of onset in the 6th decade of life [9]. Rare occurrences of atrial myxoma in a pediatric age group have also been reported [10]. Li et al. [11] evaluated the occurrence of cardiac myxomas in men and women. He found that the ratio of women to men for left atrial myxomas was 2.05:1, while that of right atrial myxomas was 0.75:1 [11]. A French study conducted by Pinede et al. [12] showed that myxomas can lead to a triad of complications, the first being obstruction (67%), followed by emboli (29%), with or without constitutional symptoms (34%).

Causes of ischemic stroke in young adults differ from those in older adults. In older adults, most of the ischemic stroke are caused by cerebrovascular atherosclerosis or cardiogenic emboli related to atrial fibrillation, cardiomyopathy or valvular disease. In young adults, most ischemic strokes are caused by hypercogulability, nonatherosclerotic arteriopathies, illicit drugs use and emboli originating from structural cardiac abnormalities [1]. Atrial myxoma, the most common benign cardiac tumor is found more commonly in young adults who present with ischemic stroke (1 in 250) than older patients with these problems (1 in 750) [13]. Primary tumors of heart are rare, with an incidence between 0.0017% to 0.19% in unselected patients at autopsy [14]. Myxomas can present in all age group but, they are particularly frequent between third and six decades of life.

Ten percent of cardiogenic embolization occurs because of mitral valve prolapse, paradoxical emboli, endocarditis, and cardiac myxoma [15]. Neurologic complications resulting from cardiac myxomas are seen in 20-35% of patients. Stroke is the initial presentation in 50% of patients with myxomas, and in 75% of patients it is seen with left atrial myxoma [16]. Calcifications on the chest radiography are more diagnostic of RA myxoma than in LA myxoma. CT & MRI can help identify the extent of the tumor and its relationships to surrounding cardiac and thoracic structures. Transesophageal echocardiography (TEE) is 100% sensitive for diagnosis of myxoma [17]. It yields morphologic detail in the evaluation of cardiac tumors, including points of tumor attachment and degree of mobility. Intraoperative TEE monitoring can aid in recognizing and avoiding tumor embolization. Cardiac MRI can assist in delineating the tumor size, attachment and mobility. This may be helpful in surgical resection, because of the risk of further emobilization; surgery cannot be deferred even in asymptomatic cases. Removal of atrial myxoma carries an operative mortality rate of 5% or less [18]. Atrial myxomas can recur following surgical resection and overall risk of recurrence is approximately 12% for familial tumors and only 1% to 3% for sporadic tumors. Neurological sequelae after resection are rare but may occur without recurrence of the cardiac tumor.

Conclusion:

- Thorough clinical examination (particularly cardiac) still remains one of the best guide to diagnosis.
- In cases of acute ischemic stroke in young adult, atrial myxoma should be included in the differential diagnosis.
- Detection of the tumor by echocardiography, followed by surgical excision, significantly reduces the risk of subsequent ischemic strokes.

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