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CASE REPORT

Takayasu's Disease: A Rear Case of Occlusive Vascular Disorder of Brain

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ABSTRACT

Takayasu's Disease is a type of inflammatory disease which involve the vessels at different levels. This disease is important for remission and relapse. It usually affects the younger age group resulting in morbidity of the most valuable group of society. Its symptoms are non specific but it may present with the involvement of different organs of the body like heart, renal impairment, fibrosis of retroperitoneal area. Diagnostic tool is Ultrasonography but definitive diagnosis is achieved with the help of CT angiography at the level of aortic arch, neck and brain. Treatment includes both medical and surgical options. Most of the time relapse occurred but with various treatment we can prolong the remission span and improve the quality of life of the patient.

KeyWords: *Takayasu's Disease, Aortic arch, Remission and relapse.*

CASE HISTORY

Mr. Shahid Bilal 27 years male R/0 Hafizabad presented in Neuro-OPD of Lahore General Hospital Lahore on 20-09-2008 with the Chief Complaints of Right sided weakness of the body for 3 months. Pain in left half of body for 2 months. Palpitations, sweating, severe weakness, headache for 24 hours. On admission patient was anxious, blood pressure was not recordable in any part of body, only left radial pulse was palpable which was of low volume very feeble and rate was 89/min. Patient was complaining about dysasthetic pain in head, heaviness. Power was 2/5 in Rt. Sided upper limb and 3/5 in right sided lower limb. Hypereflexia was present in right sided upper and lower limb, planter was up going on this side and abdominal reflexes were absent on right side of the body. Patient was admitted in hospital.

After detail history and examination, routine investigations were carried out. These included Blood R/E, Blood sugar. Blood urea Serum cretinine, LFT's. X-ray Chest P-A view and E.C.G. All the investi-zgations were negative except ESR which was above 80 mm/lst hour. As hemipresis was present therefore CT Scan was done which revealed a dense infarct in the area of Internal Capsule (Fig. 1). Vascular

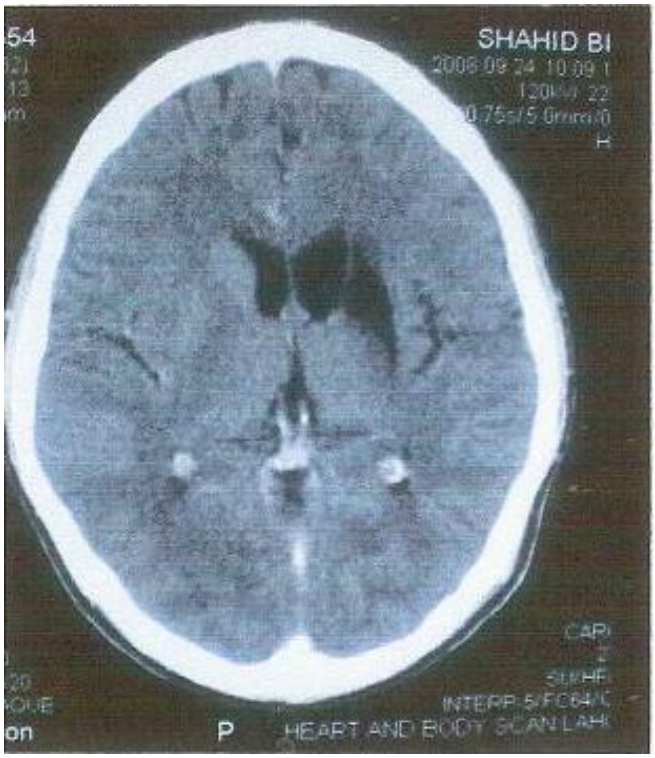

Fig. l: *C.T Scan Brain Plain Dense Infarct in left Para ventricular area.*

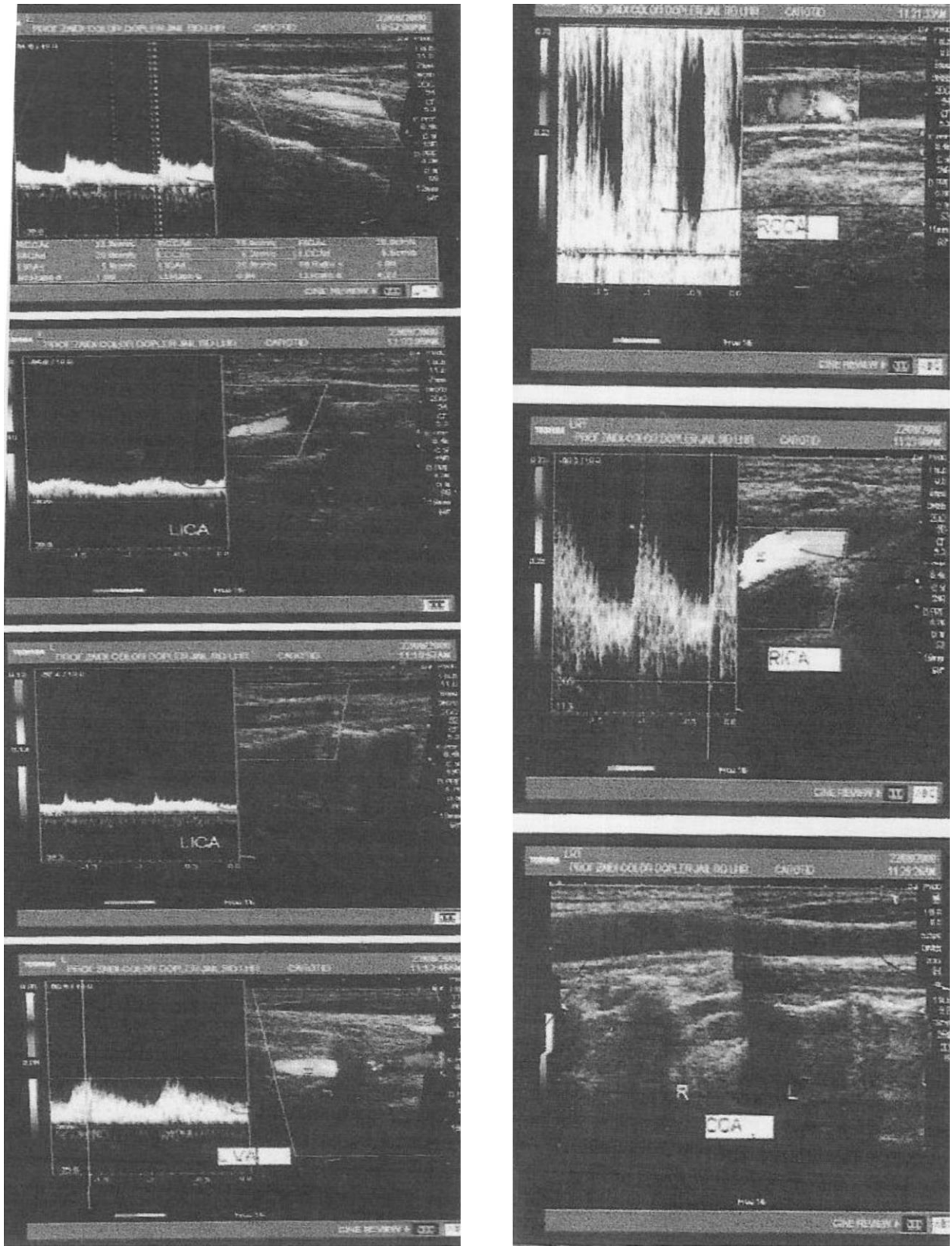

Fig. 2: *Report of Carotid Doppler Studies revealed Narrowing in different Vessels at multiple levels.*

examination of the patient, history and previous history of TIAs all were indicative some Occlusive vascular disorder of brain therefore patient was subjected to througal cardiovascular evaluation. We went for Echocardiography and Carotid dopplar studies. Echocardiography was normal but Carotid dopplar studies revealed very severe stenosis in proximal Left Common Carotid Artery (LCCA) and low Doppler signals to LCCA and ICA. Very high dopplar signals seen in Right Common Carotid Artery (RCCA) and Right ICA. While vertebral arteries were normal. (Fig. 2). Patient was referred to Punjab Institute of Cardiology where Tran thoracic imaging report revealed a normal biventricular systolic system.

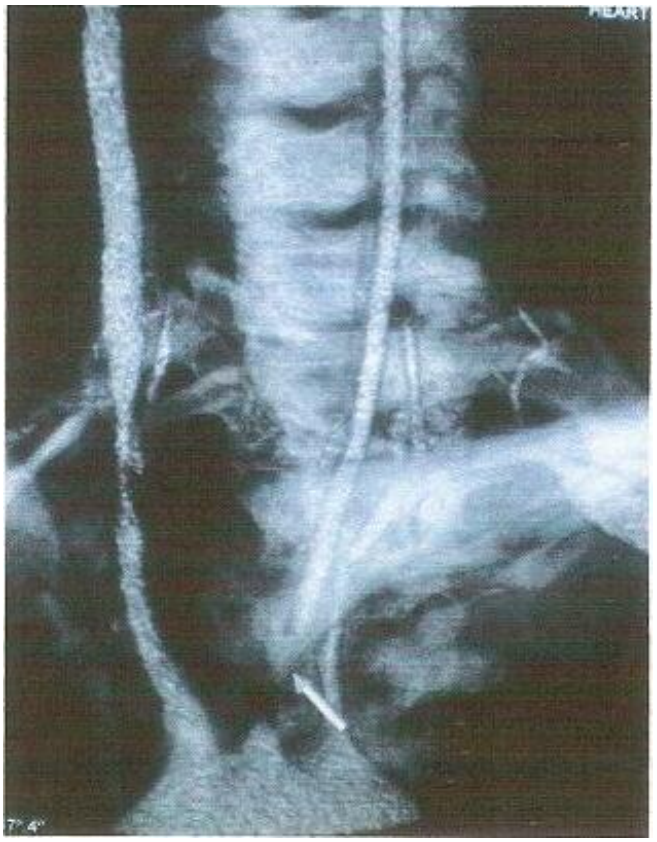

Fig. 3: *Narrowing at the origin of Left Common Carotid Artery (LCCA).*

Due to abnormal flow in carotid Doppler, patient was subjected to complete analysis of vascular system especially at the level of aortic arch, neck and brain. CT Angiography were carried out at thoracic , neck and brain levels and it revealed Narrowing of Left CCA right at the origin from thyrocervical trunk (Fig. 3). Narrowing of Right and Left Subclavian arteries at their origins (Fig. 4). Narrowing of Right Common

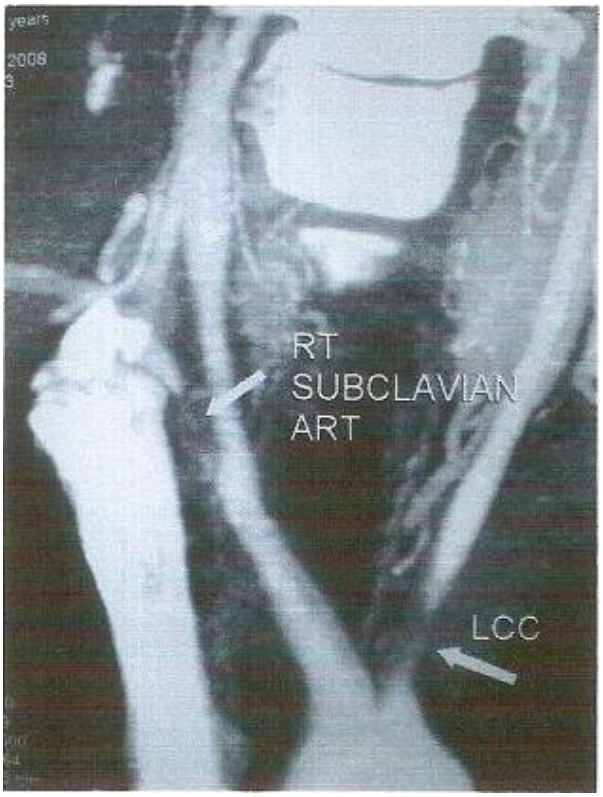

Fig. 4: *Narrowing at the level of Right Subclavian Artery.*

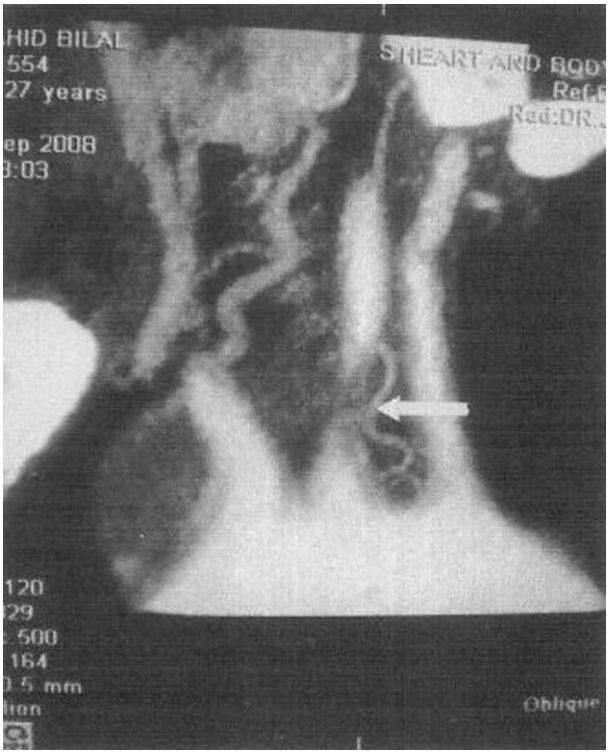

Fig. 5: *Narrowing of Right Vertebral Common Carotid Artery in Neck.*

carotid artery at neck (Fig. 5). Narrowing of Right Vertebral artery at its origin (Fig. 6). Segmental narrowing of Left Middle Cerebral Artery after bifurcation of Internal Carotid Artery intracranially (Fig. 7 and 8). Aortiris was noted in the descending part of Aorta as well. Considering the picture the final diagnosis was made i-e Takayasu's Disease. Patient was treated symptomatically with analgesics, steroids in tapering doses, antiplatelet therapy, vasodilators, microvascular insufficiency relieving medicines and cticholine. Cardiologist, Rheumatologist, medical Specialist and radiologist along with neurosurgeon examined the patient at different points and agreed on the diagnosis of Takayasu's disease. No procedure was offered by the cardiologist or vascular surgeon and it was decided to continue the conservative management for the patient. Patient remained admitted for 2 weeks and finally discharged in a better condition and with relief of symptoms. He was advised to make regular follow-up visits. Exaggeration of symptoms was noted thrice in the last one year, which was treated with tapering steroids. An episode of depression occurred in which tricyclic anti depressant was given with effective success. In the last one year excerrebation of events occur three times but now the time gap is increasing.

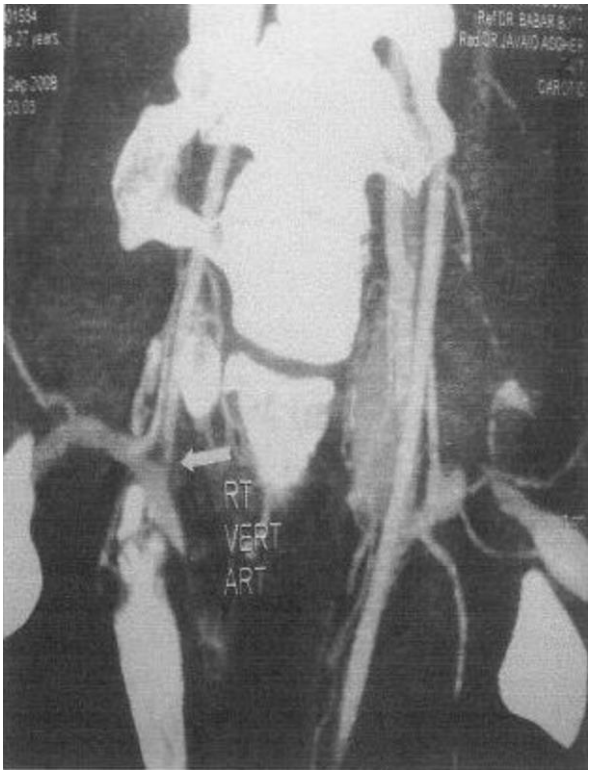

Fig. 6: *Narrowing of Right Artery at its origin.*

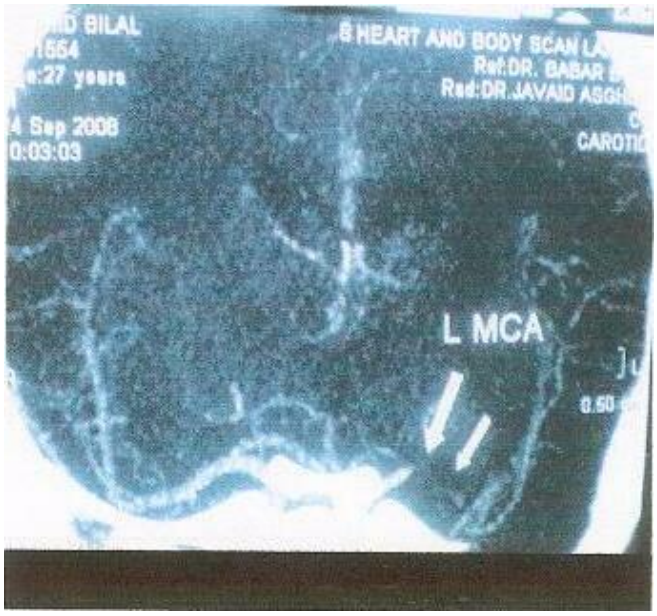

Fig. 7: *Narrowing of Left Middle Cerebral Artery (Coronal Section).*

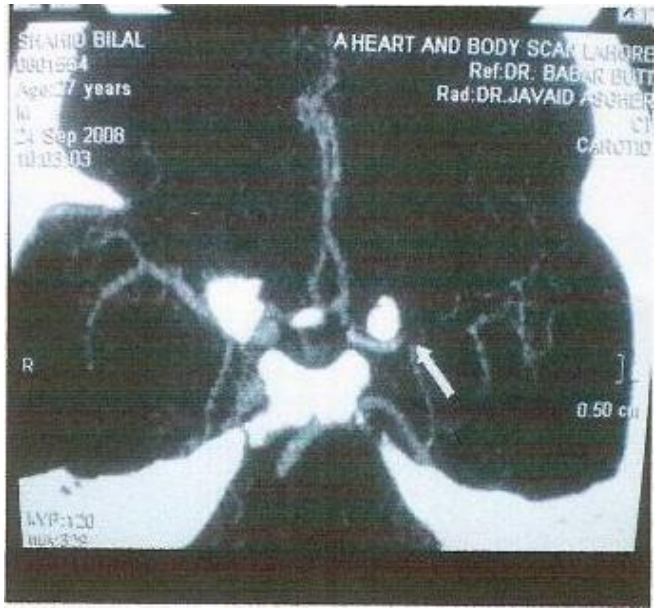

Fig. 8: *CT Angiography Left Middle Cerebral Artery (Axial Section).*

Takayasu's Disease is an inflammatory disorder[1] which is not typical intracranial vascular disease. It involves predominantly the vessels around the arortic arch[2&12] Still, in younger patients[3] presenting with symptoms of cerebrovascular event. Takayasu's disease should be considered. It occurs in all racial groups, with an unexplained prediction in Asians. A report from Olmsted County, Minnesota, indicates an

incidence of 2.6 per million per year. The female - male ratio is 9:1 and most patients present with symptoms when they are in their thirties or forties. The classic presentation of Takayasu's disease includes stenosis of arterial ostium at the origin of the aorta. Lesions can also occur at the aortic arch, subclavian artery in up to 53% cases called "pulse less disease", carotid system (30%), pulmonary or renal artery 45% and all other segments of aorta. Histopathologically an acute infiltration of lymphocytes of the entire vessel wall occurs with destruction of elastic fibers and smooth muscles cells.[4] Post inflammatory sclerosis of the vessel walls causes arteriosclerotic plaques that finally occlude the vessel lumen; saccular aneurysms may be observed.

Symptoms at disease onset are nonspecific, including fever, weight loss, fatigue, or artheralgias[5] over months. Neurological symptoms include headache (50.3%), hemiplegia (7.7%), visual disturbances (14.9%), parasthesias 940.1%) and seizures (6.3%). Unusual presentations includes pericardial effusion[6] intramural Haematoma[7] and retroperitoneal fibrosis.[8] A 5 year survival of 83% has been reported; three of eight deaths were caused by stroke. A worsening of symptoms during pregnancy may occur. No conclusive etiological concept has been developed. A correlation with tuberculosis or hereditary predictors was discussed but has not been validated.

The diagnostic evaluation is based on angiography findings for the arortic arch and the descending aorta. Duplex ultrasonography has proved to be valuable tool for the examination of the carotid system, being even more sensitive than angiography in demonstrating the typical circumferential intima and media thickening. Other than an accelerated ESR of 30-120 mm/h in most of the patients all blood tests are typically negative. No controlled treatment reports have been published so far. Most authors recommend steroids (0.5-1.5 mg/kg per day of prednisolone) and cyclophosphamide (starting with 2mg/kg per day) therapy during the inflammatory stage. Long term remission is reported and confirmed by few investigator with effective use of Infliximab on long term basis.[9] Surgical procedures in cases of stenotic or even occluded vessels include different types of by pass operations[13] Stenting[10] or reconstruction[14] arteries. These operations have been successful in most patients even on a long term basis.[11]

CONCLUSION

Takayasu's Disease is a rare entity of inflammatory vascular disorders. It affects younger age group. Sometime it presents as stroke. Complete evaluation of the vascular occlusive disorder especially in the younger age group is recommended.

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REFERENCES

1. Byme SC, Egan B, Tiemey S, Feeley M. In Takayasu's arteritis. In, Med J. 2008 Nov-Dec; 101 (10): 320. PMID19205149.
2. Ateş S, Duvan I, Kurtoğlu M, Bakkaloğlu B, Beşbaş S, Aybek T. Involve sparing ascending and transverse aorta aneurysm repair with mild hypothermia in Takayasu arteritis. Anadolu Kardiyol Derg. 2009 Feb; 9 (l): 73. PMID: 19196587.
3. Maffei S, Di Renzo M, Bova G, Auteri A, Pasqui AL. In: Takayasu's arteritis: a review of the literature. Intern Emerg Med. 2006; 1 (2): 105-12.
4. Deng J, Ma-Krupa W, Gewirtz AT, Younge BR, Goronzy JJ, Weyand CM. In, Toll-like receptors 4 and 5 induce distinct types of vasculitis. Circ Res. 2009 Feb 27; 104 (4): 488-95. Epub 2009 Jan 15.
5. Pariser KM.: Takayasu's arteritis. Curr Opin Cardiol. 1994 Sep; 9 (5): 575-80.
6. Fateh-Moghadam S, Huehns S, Schmidt WA, Dietz R, Bocksch W. In: Pericardial effusion as primary manifestation of Takayasu arteritis.hit J Cardiol. 2009 Feb 2. [Epub ahead of print] PMID: 19193462.
7. Marla R, Migrino R, Osipov V, Lilly RE.In, Takayasu arteritis presenting as Type A intramural hematoma - An unusual presentation. Hit J Cardiol. 2009 Jan 28. [Epub ahead of printj PMID: 19185361.
8. Jghaimi F, Kabbaj A, Essaadouni L. In: akayasu's arteritis and retroperitoneal fibrosis: A case report. Rev Med Interne. 2009 Feb 19. [Epub ahead of print] French. PMID: 19231040.
9. Krivosheev OG, Smitienko IO, Aslanidi IP, Mukhortova OV. In: Persistent remission of Takayasu aortoarteritis induced by long-term treatment with infliximab and confirmed by repeat positron emission tomography. Ter Arkh. 2008; 80 (10): 90-3.
10. Us M, Numan F, Goksel OS, Basaran M, Yihnaz AT. M, Stenting for stenosing Takayasu aortitis following carotid artery stenosis in a 32-year-old patient. Vascular. 2008 Sep-Oct; 16 (5): 283-6. PMID: 19238871.

11. Ogino H, Matsuda H, Minatoya K, Sasaki H, Tanaka H, Matsumura Y, Ishibashi-Ueda H, Kobayashi J, Yagihara T, Kitamura S. In: 0verview of late outcome of medical and surgical treatment for Takayasu arteritis. circulation. 2008 Dec 16; 118 (25): 2738-47. Review. PMID: 19106398.
12. Melo NC, Sette LH, Coelho FO, Lima-Verde EM, Santana AN, Praxedes JN. In: Clinical images: Hypertension due to otherwise asymptomatic, complete aortic occlusion in Takayasu arteritis. Arthritis Rheum. 2009 Jan; 60 (l): 312. PMID: 19116925.
13. Hiu T, Kitagawa N, Suyama K, Nagata LIn, Progressing takayasu arteritis successfully treated by common carotid-internal carotid crossover bypass grafting: technical case report. Neurosurgery. 2008 May; 62 (5): E1178-9; discussion El 179.
14. Kawaguchi T, Fujita S, Ijichi A, Shose Y, Nakamura E, Mori E. In: Successful reconstruction of completely obstructed right CCA in Takayasu's disease: usefulness of rapid sequence scanning method—case report. Neurol Med Chir (Tokyo). 1990 Dec; 30 (13): 1024-8.