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Case Report

A RARE CASE OF ONE AND HALF SYNDROME AFTER SURGICAL CLOSURE OF ATRIAL SEPTAL DEFECT

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ABSTRACT

We report a 17 year old girl who presented to us with atrial septal defect (ASD) of Osteum secundum (OS) type which was operated by ASD patch closure using double velour dacrone (DVD). There was no evidence of intracardiac thrombus preoperatively confirmed by transesophageal echocardiography and as directly visualized during intraoperatively. She developed left hemiparesis with right sided horizontal gaze palsy and internuclear opthalmoplegia (one and half syndrome) 48 hours after the operation. Though de-airing was done during procedure, air embolism was suspected as probable mechanism for neurological deficit in our case as there was no evidence of intracardiac thrombus. Cerebral air embolism following cardiothoracic surgeries known to cause neurological manifestations, but they are in the form of altered sensorium commonly, followed by hemiplegia/monoplegia. One and half syndrome following cardiothoracic procedures been mentioned in literature, but there are no case reports following ASD closure and following cerebral air embolism.

Key Words: *One and a Half Syndrome, Air Embolism, Paramedian Pontine Reticular Formation, Internuclear Opthalmoplegia*

INTRODUCTION

One and half syndrome (OAHS) is an ocular movement disorder characterized by lateral gaze palsy to one direction associated with internuclear opthalmoplegia (INO) to the opposite direction was first described by miller fisher in 1967. The syndrome usually results from a single, unilateral and relatively small lesion at dorsal tegmentum of the lower pons (Fisher, 1967). Vascular and demyelinating are important causes for OAHS but neoplastic disorders also known to cause OAHS (Maranhao *et al.*, 1996). Cerebral air embolism following cardiothoracic and neurosurgical procedures are known to cause neurological manifestations in the form of altered sensorium more commonly (Jain, 1996). Focal neurological deficits such as hemiplegia or monoplegia may occur following air embolism (Jain, 1996).

CASES

A 17 year old girl presented with history of recurrent chest infection and poor health since birth. Since last 6 months she experienced progressively increasing exertional dyspnea, palpitation and chest pain. Dyspnea was associated with cough which was non productive in nature. At the time of admission she had pallor and on auscultation fixed wide split of second heart sound with ejection systolic murmur in pulmonary area. Her transesophageal echocardiography (TEE) showed Atrial septal defect (ASD) of osteum secundum (OS) type and there was no evidence of any thrombus in atria.

ASD closure was done by Double velour dacron (DVD) patch under cardiopulmonary bypass in arrested heart. Right atrium was opened and interatrial septal defect of size 4x4 cm was closed with DVD patch. Routine de-airing of the heart was done before declamping the aorta and allowed heart to beat. Carbondioxide (CO_2) insufflations though superior, were not done in our patient but proper de-airing was done to prevent air trapping in cardiac chamber. No evidence of atrial thrombus was found when heart was opened up during the procedure and coronary sinus was found normal. Operation was uneventful and

was shifted to intensive care unit (ICU). She was weaned off from ventilator eventually and extubated next day morning and she did not develop any new signs or symptoms. Routine transthoracic echocardiography (TTE) was done after 24 hours showed DVD device in situ.

However 48 hours after the surgery it was noticed that she had left hemiparesis. There was no history of seizures. On examination patient was conscious, oriented and had slurred speech. Her blood pressure was 110/70 mmHg and pulse rate was 90/min, good volume, regular rhythm and all the peripheral pulses were well felt. Detailed neurological examination revealed horizontal gaze palsy on looking towards right side and impaired adduction in the right eye with nystagmus on left eye abduction typically suggestive of one and half syndrome. She had normal vertical eye movements and convergence. She also had upper motor neuron type of left facial paresis with facial deviation towards right side and labial dysarthria. She also had left hemiparesis with power of 3/5 in upper limb, 2/5 in lower limb with brisk reflexes and extensor response of left plantar. Her sensory examination was normal.

Her blood investigations revealed haemoglobin of 9.8%, peripheral smear showed microcytic hypochromic anemia and rest of the routine investigations were normal. Her brain MRI on T1 weighted image showed hypointensities in right parapontine region including tegmentum (Fig. 1A) with corresponding hyperintensities on T2 weighted and T2 Flair images (Fig. 1B). Diffusion weighted images revealed restricted diffusion suggestive of acute infarct in right parapontine region. Her ECG was normal without any ST-T changes, atrial fibrillation, atrial tachy arrhythmias. Repeat TEE showed DVD device in situ and there was no evidence of atrial aneurysm or thrombus in the atria. Cerebral air embolism was suspected in our case though transcranial Doppler (TCD) was not done as it is not available in our institution. With high index of suspicion of cerebral air embolism, she was treated with 100 % oxygen using closely fitting mask within 6 hours, injectable steroids (dexamethasone) and aspirin. Hyperbaric oxygen per say could not be administerted as hyperbaric chamber facilty is not available in our institution. Antiepileptic drugs were used prophylactically. Her power improved over a week to 4/5 in upper limb and 3/5 in lower limb and slight improvement in her gaze. She was discharged and followed up thereafter on aspirin treatment. One month later on follow-up, patient's extraocular movements were complete and there was no gaze restriction.

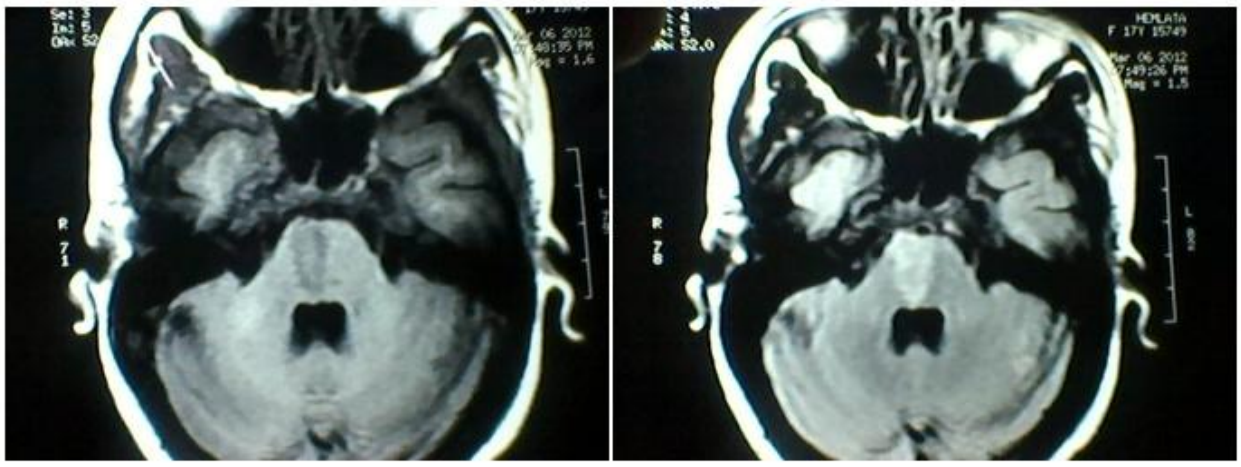

Figure 1: A-T1 Weighted MRI image showing hypointensities in right paramedian pontine region including tegmentum. **B**-T2 Flair image showing hyperintensities in corresponding areas

DISCUSSION

One and half syndrome is characterized by impaired abduction and adduction in one eye and impaired adduction with nystagmus upon abduction in another eye. It constitutes conjugate lateral palsy in one direction, plus one half of gaze palsy in the other (Fisher, 1967). Structures involved in one and half

syndrome are parapontine reticular fibres (PPRF) responsible for horizontal gaze palsy and ipsilateral median longitudinal fasiculus (MLF) responsible for impaired adduction due to INO (Maranhao *et al.*, 1996). A medial pontine lesion can affect both the PPRF on one side and the MLF crossing from the contralateral side. Because of the ipsilateral PPRF lesion the patient will have gaze palsy to the same side and because of the MLF lesion the patient will have INO on the same side.

Multiple sclerosis and brain stem infarction are most common causes of one and half syndrome. Less frequently it is caused by primary and metastatic tumors of the brainstem and cerebellum (Newton and Miner, 1991), even brainstem hematomas/bleeding are causes of OAHS. Rarely one and half syndrome can develop postoperatively after the removal of tumours of the posterior fossa. Newton and Miner, 1991 reported a case of one and half syndrome after a resection of a midline cerebellar astrocytoma. Other than removal of local tumor and metastasis remote surgeries has been rarely been mentioned as a cause of OAHS. Cerebral air embolism has not been mentioned as a cause for OAHS so far. Though neurological sequelae have been estimated to occur in 19-50% of the patients with cerebral air embolism (Jain, 1996), sudden change in sensorium is the most common symptom and ranges from disorientation to coma. Hemiplegia or monoplegia can occur.

The most common reason for air to enter the arterial system during extracorporeal perfusion is that the blood level inside the oxygenator gets too low. Clinical manifestations depends on the patient's posture, the route of entry of air, the volume of air, the size of the bubbles and the rate of entry of air (Jain, 1996). Arterial air embolism is known following cardiothoracic and neurosurgical procedures. Massive arterial air embolism is a very rare complication during extracorporeal circulation. The lung is usually an effective filter for air bubbles. The introduction of air into the venous or arterial system can cause air embolism leading to severe neurological deficits. The most common cause reported in the literature is iatrogenic. The incidence of air embolism during cardiopulmonary bypass operations is 0.1% and air enters venous system in 30- 40% of the patients undergoing neurosurgical operation. In iatrogenic cases the air is either sucked into the veins with negative pressure or introduced into the veins or arteries under pressure (Jain, 1996).

Air embolism also can occur due to inadequate de-airing before declamping of aorta and allowing heart to beat especially when CO_2 carbon dioxide insufflations is not used during open heart surgery in operating field which is considered as ideal method to prevent air embolism. CO_2 field flooding is more effective than mechanical de-airing in removing intracardiac bubbles following valvular surgery (Kalpokas *et al.*, 2003). In present case air embolism has occurred despite adequate de-airing protocol and was presented late in postoperative period after 48 hours gives suspicion that probably air got trapped in left atrium and got embolised later during change of posture. Careful standard cardiac de-airing did not prevent air embolism caused by the delayed release of air trapped in the lung vessels. Though not done in our case routine use of intraoperative transesophageal echocardiography (ITEE) is recommended to assess the thoroughness of de-airing procedures (Tingleff *et al.*, 1995). This will help eliminate air embolism or at least lead to an increased awareness of the problem of retained air. Minimizing air embolism during open heart operations should contribute to a reduction in central nervous system damage and improvement of intellectual function after heart operations (Tingleff *et al.*, 1995).

TCD studies show that microscopic cerebral artery air emboli (CAAE) are present in virtually all patients undergoing cardiac surgery[3]. Treatment measures include administration of hyperbaric oxygen (HBO) (Jain, 1996). In some cases diagnosis is proven only after successful response to hyperbaric therapy. A bolus dose of steroid (10mg dexamethasone) can be given to prevent cerebral edema (Jain, 1996). Antiplatelet drugs have been used and the use of heparin as anticoagulant is considered as risky but if patients who are already on heparin have better prognosis after air embolism than those who are not anticoagulated (Jain, 1996). An antiepileptic medication may be required for control of seizures and one can use prophylactically. In our patient though there was no evidence of cerebral edema bolus dose of injectable steroid was given. Heparin was not used in our patient but aspirin was given during the hospital stay and after the discharge in follow up. Though our patient received 100% oxygen through high flow

mask along with steroid and antiplatelet drug, possibility of spontaneous recovery as a natural course cannot be ruled out.

However one and half syndrome following a cardiothoracic surgery or interventions been rarely reported. Anderson *et al.* reported a rare case of one and half syndrome with supranuclear facial palsy following transluminal coronary angioplasty (Anderson *et al.*, 1999). Neurological manifestation following ASD closure also been rarely reported. Huber et al reported case of 5 year old girl who suffered massive air embolism during surgical closure of ASD (Huber *et al.*, 2000). The neurologic examination in that patient showed pathological signs in the form of spontaneous oral twitches and asymmetric movements of the extremities with weakness mainly on the left side.

To conclude OAHS following surgical closure of ASD/ and or following cerebral air embolism has not been reported so far to the best of our knowledge. We report a case of OAHS with left hemiparesis following surgical closure of ASD. Probable mechanism of stroke in our patient was air embolism as DVD device was in situ and there was no evidence of thrombus in atria by TEE preoperatively and post operatively, when heart was opened intraoperatively, and as there was no hypotensive episodes perioperatively. Though not done in our patient, ITEE should be done whenever possible, TCD and treatment with HBO should be considered whenever facilities are available. A high index of suspicion is very important in diagnosis. Because under suspicious circumstances air embolism should be assumed present unless otherwise proven.

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