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A CASE OF PRIMARY PULMONARY HYPERTENSION WITH aCL POSITIVE

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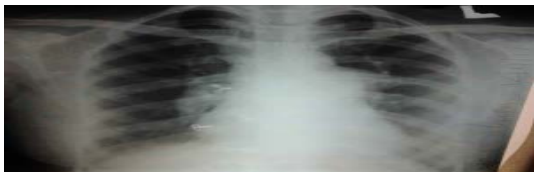
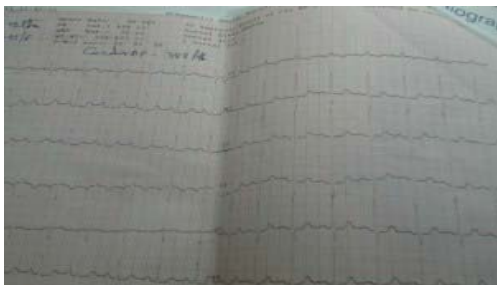
Abstract : ABSTRACT- Primary pulmonary hypertension is a diagnosis of exclusion. we present a case with aCL positive but patient not having any thromboembolic manifestations. Its a rare entity ,but if diagnosed early can be treated and prognosis is good.

Keyword :KEYWORDS- Primary pulmonary hypertension, aCL positive

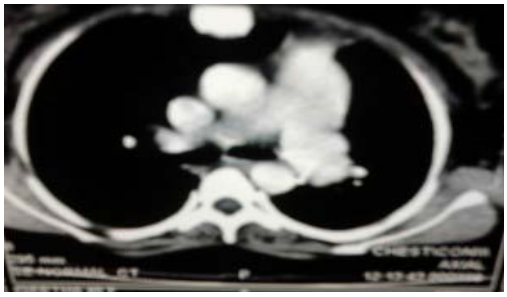
CASE HISTORY: Patient aged 50 years presented with complaints of chest pain 3 months and breathelessness 3 months, which was grade lll in nature, and palpitation on and off. No other specific complaints. Past history patient had TB, at 5yrs of age and treated . On examination patient CVS s1s2 normal .palpable p2 present and loud p2 .other system normal. patient was suspected of pulmonary hypertension and investigated. Blood investigations- CBC-TC9050,-DC- P-56%,L-42%E- 2%. Hb-10.5 ,esr-90/130. Platelet count 2 lakhs. Urine routine normal. Chest x ray- cardiomegaly with right pulmonary artery dilated. Pulmonary function test – moderte restriction. CT chest- pruning of pulmonary arteries +with features suggestive of pulmonary hypertension. Portal doppler- normal study. Echo- RA/RV dilated, TRpg-4/64 ,septum intact ,severe pulmonary hypertension Ecg- RVH with strain pattern Anti ds dna, anti ssA and ssB , scl70 anti smith ab,antiJo ab, RNP smith antibody, aPL all negative, aCL-POSITIVE, complement c3-115, c4-30.6, prothrombintime-24.6,INR- 2.39.CTangio of chest- 40%LAD stenosis, Barium swallow- wide pulmonary bay, desending branch of right and left pulmonary artery no significant abnormality. Other tests Thyroid function, HIV,USG all nomal study. So patient was diagnosed as primary pulmonary hypertension with aCL positive and treated with Sildenafil 25 mg bd, warfarin 2mg od and titrated according to INR, Enalapril 2.5 mg bd, Deriphylline 100 mg bd. Patient improved well and was maintained on these drugs. **x ray**



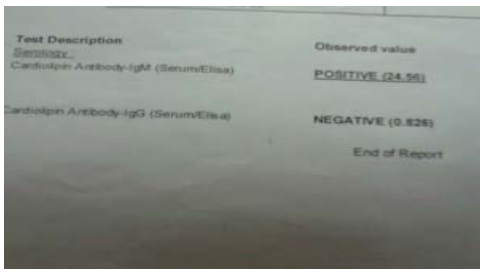
x ray chest- right pulmonary aretry dilated



ecg- right ventricular hypertrophy



ct chest



DISSCUSSION

Pulmonary hypertension is abnormal elevation in pulmonary artery pressure. Idoipathic pulmonary hypertension is a name given to pulmonary hypertension of unexplained cause. Its incidence two per million.More common in women. The prevalence of pulmonary hypertension in APS associated with SLE and the primary APS has been estimated to be between 1.8% and 3.5%, respectively.1The association of pulmonary hypertension with aPL was first reported in 1983.2 Several cases of primary pulmonary hypertension complicating primary APS have been described. 2345The pathophysiology of IPAH is poorly understood. An insult (hormonal, mechanical, other) to the endothelium may occur, possibly in the setting of increased susceptibility to pulmonary vascular injury (multiple hit theory), resulting in a cascade of events characterized by vascular scarring, endothelial dysfunction, and intimal and medial (smooth muscle) proliferation. At least 15-20% of patients previously thought to have IPAH actually have a familial form. The most common genetic defect in these cases involves the BMPR-II gene.However only about a third of affected patients witha family history of PAH have an identifiable BMPR-II mutation. The most common symptoms and their frequency, reported in a national prospective study, are as follows: Dyspnea (60% of patients) Weakness (19%) Recurrent syncope (13%) Women are more likely to be symptomatic than men. On examination - second heart sound is usually increased, which may demonstrate fixed or paradoxic splitting in the presence of severe right ventricular dysfunction; occasionally, the second heart sound may be palpable. Pulmonic regurgitation (Graham Steell murmur) may be apparent. Murmur of tricuspid regurgitation can be present, and a right ventricular lift (heave) may be noted.Jugular venous pulsations may be elevated in the presence of volume overload, right ventricular failure, or both, large waves due to tricuspid regurgitation. Investigations include routin e blood inestigations, chest xray, ecg, ct chest, pulmonary angiography, dlco, thyroid function test and serology test for connective tissue disorders. To manage pulmonary hypertension, chronic anticoagulation is needed in all cases to prevent the occurrence of new thrombotic event .In some patients with very severe disease successful thromboendaterectomy has been performed.789vasodilators such as calcium channel blockers are used10only one third of patients respond to this vasodilator challenge.10,11 other drugs used for treating are endothelin receptor antagonist,bosenton,ambrisentan12,13.Phosphodiesterase inhibitor sildenafil,tadalafil.14,15Oral Prostacyclins16,17and iv prostacyclins are used.18,19Lung transplantation is the final treatment if medical therapy don't work.20.

CONCLUSION- Though primary pulmonary hypertension with acl positive and no thromboembolic manifestations is rare entity it can be easily treated if promptly diagnosed and patient prognosis is better if treated early.

REFERENCES

1.Kamashta MA, Cervera R, López-Soto A, et al. Comparison of the primary and secondary antiphospholipid syndrome

2. Mackworth-Young CG, Boey ML, Hull RG, et al. Pulmonary hypertension in systemic lupus erythematosus 3.Ashreson RA, Lahita RG Primary idiopathic pulmonary hypertension complicated by pulmonaryarterialthrombosis.

4. Yasuma K, Katsuki T, , Nakamura Y, et al. Primary antiphospholipid syndrome and pulmonary hypertension with prolonged survival. A case report.

5. Martínez-Guerra ML, et al. Treatment with thromboendarterectomy. De la Mata J, Gomez-Sanchez MA, Aranzana M, Gomez-Reino JJ. Longterm iloprost infusion therapy for severe pulmonary hypertension in patients with connective tissue disease.

6.Haworth SG. Primary pulmonary hypertension. J R Coll Physicians Lond1998;32:187.

7.Cucurul, L, Orriols R, Vilardell M. Tromboendarterectomía pulmonar en un paciente con

síndrome antifosfolípido primario. Med Clinc.

8. Sandoval J, Amigo MC, Barragan R, Izaguirre R, Reyes PA, Martínez

Guerra ML, et al. Treatment with thromboendarterectomy

9.Anderson EN, Ali MR. The lupus anticoagulant, pulmonary thromboembolism and fatal pulmonary hypertension. Ann Rheum .

10.Tam L-S, Li EK. Successful treatment with immunosupression in SLE associated with secondary antiphospholipid syndrome.

11.Rich S, Kaufmann E, Levy PS. The effect of high doses of calcium-channel blockers on survival in primary pulmonary hypertension. N Engl J Med.

12.Galiè N, Hinderliter AL, , Simonneau G, Pulido T, et al. Effects of the oral endothelinreceptor on echocardiographic and doppler measures in patients with pulmonary arterial

hypertension. J Am Coll Cardiol.

13Oudiz RJ, Galiè N, Frost A, Ghofrani HA, et al. Long-term ambrisentan therapy for the treatment of pulmonary arterial hypertension. J Am Coll Cardiol. .

14.Simonneau G, Rubin LJ, FleminTR, Frost AE, et al. Addition of sildenafil to long-term intravenous epoprostenol therapy randomized trial. Ann Intern Med.

15.Galiè N, Brundage BH, Ghofran, Safdar Z, et al. Tadalafil therapy for pulmonary arterial hypertension. Circulation. [Medline].

16.Oudiz RJ, Farber HW. Dosing considerations in the use of intravenous prostanoids in pulmonary arterial hypertension: an experience-based review.

17.Shapiro SM, , Georgiou D, et al. Primary pulmonary hypertension: improved long-term effects and survival with continuous intravenous epoprostenol infusion. J Am Coll Cardiol.

18.Oudiz RJ, Rubin LJ, et al. Treprostinil, a prostacyclin analogue, in pulmonary arterial hypertension associated with connective tissue disease. Chest. . [Medline].

19.Simonneau Bourge RC, et al. Continuous subcutaneous infusion of treprostinil, double-blind, randomized, trial. Am J Respir Crit Care Med

20.AshersonRA,HigenbottamTW, Pulmonary hypertension in a lupus clinic expierence with twenty four patients.

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