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Case Report

Recurrent Pulmonary Arteriovenous Malformation with Embolization

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Abstract

Pulmonary arteriovenous malformations in majority of cases are congenital in origin. In our center we found a case, history of dyspnoea on exertion of long time, a short duration of malena and epistaxis. On clinical examination, we assumed pulmonary embolism and investigated with Computed Tomography (CT) Thorax and Pulmonary angiogram. Our findings were faint bruit over right mammary area and diagnosis was pulmonary arterial malformation. Patient was successfully treated by embolotherapy using steel coils. After couple of week's patient returned with dyspnoea, small Pulmonary Arteriovenous Malformation (PAVM) was again identified and repeated embolotherapy. Result showed in 90% exclusion of the aneurysm from the rest of the systemic circulation.

Keywords: Recurrent pulmonary arteriovenous malformation; Embolotherapy; 90% Exclusion

Introduction

PAVM is a rare disease with abnormal symptoms between the pulmonary arteries and pulmonary veins. The etiology of this disease is usually congenital, but they required various conditions, such as mitral stenosis, schistosomiasis, tuberculosis, trauma and metastatic thyroid carcinoma among others.

The Hereditary Hemorrhagic Telangiectasia (HHT) also known as Osler-Weber-Rendu Syndrome (OWRS) [1]. OWRS is a genetic disease, an autosomal dominant disorder characterized by telangiectasias and aneurysms with primary involvement of the mucosa. The reported prevalence of this disease is 1 to 2 cases per 100 000 population [2]. The disease most commonly occurs in white patients, few studies have reported in Asian and African countries, and most frequently after 30 years age group and equal severity in both genders but may be clinically silent and majority of cases of OWRS with pulmonary arteriovenous malformation treated successfully with pulmonary arterial embolization using steel coils [2-4].

Case Presentation

A 40 years old female patient presented with breathlessness since 8 years with recent intermittent non-pleuritic chest pain. On examination patient have hemodynamically stable with a pulse rate of 90 per minute, blood pressure of 140/90 mm of Hg and oxygen saturation of 90% on finger held pulse oximetry. Initial Respiratory system examination had normal air entry bilaterally with vesicular breath sounds. Cardiovascular, abdominal and neurological examinations were normal. Investigations showed haemoglobin of 12.1 gm/dl, packed cell volume of 56%; white blood cell count of 9,400 per cubic mm, (polymorphs 79%, lymphocytes 17%, eosinophils 2% and basophils 2%). Other biochemical parameters were within normal limits. Electrocardiogram (ECG) showed sinus tachycardia with right axis deviation and R wave in V1. Patient has few episodes of epistaxis and suspicion of pulmonary embolism, patient underwent for Computed Tomography Thorax and Pulmonary angiogram. The CT scan showed intensely enhancing soft tissue density lesion in the right middle lobe without calcification. On examination of oral cavity showed oral telangiectasias and on auscultation of right mammary and right intra scapular area, faint bruit was heard (Figure 1A).

Pulmonary angiography noted pseudoaneurysm with arteriovenous fistula arising from the right pulmonary arterial branches (Figure 1B). We performed steel coiling Post-coiling angiogram revealed 90% exclusion of the pseudoaneurysm from the circulation (Figure 2). Post-procedure the patient was symptomatically improved. The arterial blood gas analysis showed a pH of 7.430, PCO2 - 38.5, PO2 - 64.7, SaO2 - 87.3 and HCO3 - 25. The patient remained asymptomatic for 2 weeks following embolotherapy. Recurrence of

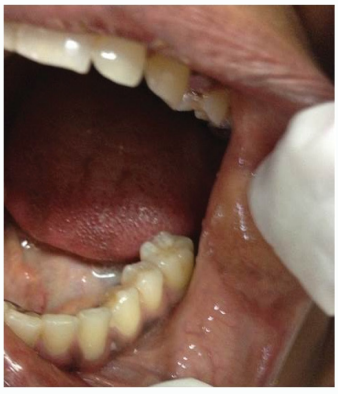

Figure 1A: Oral telangiectasias.

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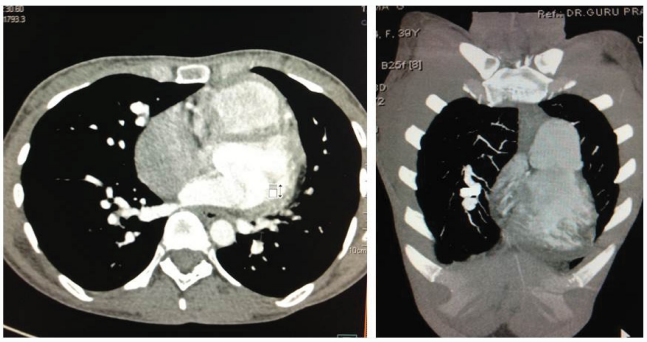

Figure 1B: Pulmonary arteriovenous malformation.

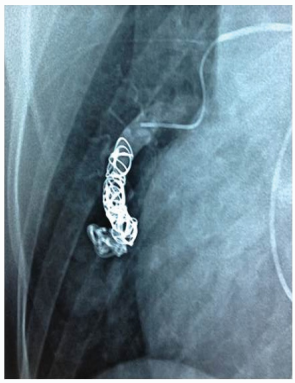

Figure 2: Post-coiling 90% exclusion of the pseudoaneurysm from the circulation.

exertional dyspnoea and epistaxis due to second AVM was noted and embolotherapy was repeated. The patient now remains asymptomatic 6 months following embolotherapy. After six months embolotherapy, patient is fine.

Discussion

Osler-Weber-Rendu is an autosomal dominant disease, which may affect small portion of blood vessels at skin surface, mucosa and organs [5]. Clinical manifestation is nasal bleeding, skin telangiectasia, headache or bruits in cerebral AVM, chest pain or dyspnea in PAVM, malena or hematochezia in gastrointestinal mucosal telangiectasia [6]. The diagnosis of HHT is made clinically on the basis of the Curaçao criteria [7]. In our case, PAVM with aneurysm formation in his right lower lung, and history of exertional dyspnoea with several episodes of nasal bleeding, in additionally, telangiectases were found on her oral mucosa. Based on the above findings our patient fit for HHT diagnosis [7,8]. A similar case reports noted by others, previous studies have found 70% to 90% of patients with PAVM were associated with HHT, and again around 30% of patients with HHT have PAVMs.2, the current case showed PAVM association with HHT [5].

The treatment options were surgical and nonsurgical procedures, patients with associated symptoms or a lesion <2 cm, were indicated for surgical procedure, the present case also noted lesion <2 cm. The first surgical procedure was performed for pneumonectomy in 1942 [5,9]. Taylor et al. [10] embolotherapy has become the major treatment options for PAVMs using balloon or metallic coils. Pollak et al. [11] reported a case series 98% success rate of the embolotherapy, in our case also we did embolotherapy. In our case recurrence PAVM present, after second embolization 90% of recanalization, a similar case reported by Kasai et al. [12], and study have established recurrence of PAVM is a common complication after embolization [13]. On expert persons performed embolotherapy the complications were less than 1% (including migration of embolization devices, symptomatic lung infarction, cardiac arrhythmia, air embolism, deep venous thrombosis transient angina pneumothorax) [14]. However in our case doesn't have any complications.

Conclusion

Osler-Weber-Rendu Syndrome is a rare genetic disease with irregular symptoms. Diagnosis is key role for treatment of either surgical or non-surgical procedures. The surgical treatment options are Embolization or silicone balloon tamponade. In generally majority of cases embolization was performed.

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