**INTRODUCTION**

Klippel-Trenaunay syndrome (KTS) is a rare congenital syndrome which involves a constellation of clinical findings and includes enlargement of veins and arteries, hypertrophy of the limb, and capillary malformations.\(^1\,^2\) KTS is seldom seen in routine clinical practice and correctly recognized by the clinicians.

The importance of this case report lies in the fact that it gives the clinicians an opportunity to recognize and understand a very rare syndrome. It also illustrates a very rare association of this syndrome which has never been described earlier and might help the clinicians, dealing with such cases, to be more alert to its presence enabling early recognition and appropriate treatment.

**CASE REPORT**

We are presenting an interesting case of a 23-year patient who presented with an episode of acute pulmonary embolism after having undergone elective laser surgery for varicose veins (Figure 1a) at another center and developed an episode of acute onset on shortness of breath with severe hypoxemia, a resting saturation of 70% along with sinus tachycardia and hypotension, and was diagnosed to have large pulmonary embolism involving bilateral pulmonary arteries. Clinically the patient had hypertrophy of right lower limb and extensive varicosities involving the limb (Figures 1b and 1c). The patient had bony deformities of the left hand and also had a bony prominence of the left forehead as a manifestation of localized gigantism, which is a part of the characteristic of KTS syndrome complex (Figure 1d). Although the presence of a port wine stain forms a part of the characteristic triad of this syndrome, but this patient had no such stain or a history of the same.

The patient was administered with Tenecteplase as a bolus fibrinolytic therapy. His symptoms improved with bolus of fibrinolytic therapy. A repeat computed tomography (CT) pulmonary angiography, to document the extent of improvement in terms of the thrombus burden, was not done although the patient still had a resting desaturation of around 87%.

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**ABSTRACT**

Klippel-Trenaunay syndrome is a rarely encountered congenital disease characterized by a triad of enlarged arteries and veins, limb hypertrophy and capillary malformations. We are presenting an interesting case of a 23-year male who had been previously diagnosed to have Klippel-Trenaunay syndrome. The patient presented with large pulmonary embolism after having undergone laser surgery for varicose veins. The diagnostic chest computed tomography (CT) performed also revealed the co-existence of severe destructive pulmonary parenchymal disease involving large areas of the pulmonary parenchyma and formation of large emphysematous bullae having an asymmetric involvement of the left lung field. The patient was managed with thrombolysis with a bolus fibrinolytic agent and subsequently underwent an inferior vena cava (IVC) filter implantation to prevent further episodes of embolism in the presence of a compromised lung. The association of Klippel-Trenaunay syndrome with recurrent pulmonary embolism and unexplained pulmonary parenchymal disease leading to almost complete destruction of large areas of lung fields is interesting and has never been previously described.

**Key Words:** Klippel-Trenaunay syndrome, Pulmonary parenchymal disease, Pulmonary embolism, Thrombolysis.
The patient was put on anticoagulation after overlapping with low molecular weight heparin, and was discharged after 3 - 4 days of admission for pulmonary embolism. He was apparently stable for 2 - 3 days following which he was referred to us for cardiac evaluation to account for the cause and to help in further management of the resting desaturation to a level of 84% - 87%, which was associated with tachycardia but no hypotension.

The patient was evaluated at our center with an echocardiogram and venous Doppler tests. The former revealed no obvious clot in the right sided chambers or in the proximal pulmonary arterial bed. There was mild dilatation of right atrium and right ventricle and the right ventricular function was normal (Figure 2a). The changes of right ventricular dilatation were significantly less in the echocardiography, performed at our institute.

![Figure 2 (a,b,c,d):](image)

**Figure 2 (a,b,c,d):** (a) Echocardiogram of the patient in subcostal four chambered view showing mildly dilated right sided chambers. (b) X-ray of the patient showing interstitial involvement of bilateral lung fields with emphysematous bullae formation of the lung parenchyma. (c) Film from the high resolution computed tomography (HRCT) of the patient showing destroyed lung parenchyma with involvement of the lung field bilaterally and bullous transformation predominantly involving the left lung field. (d) Pulmonary angiogram on the CT showing evidence of thrombus in bilateral pulmonary arteries.

The evaluation of the patient at our center was performed after the thrombolysis performed outside. The trans-thoracic window was extremely poor due to the presence of emphysematous lung parenchyma and so only a subcostal window was available from which most of the echocardiographic assessment had to be performed. The assessment of the pulmonary parenchymal was done with the help of CT and the X-ray and this revealed extensive destruction of bilateral lung fields with diffuse emphysematous bullae, predominantly involving the left sided lung field with early changes of the right side (Figures 2b and 2c). The CT angiogram also confirmed the presence of bilateral thrombus in the pulmonary arteries (Figure 2d). There was no evidence of pneumothorax but there was extensive fibrosis which had caused the mediastinum to shift to the right side. The venous Doppler of the affected extremity revealed the presence of dilated and tortuous varicose veins with evidence of thrombus in them. The serum alpha-1 antitrypsin levels were within normal range. The systemic inflammatory markers in the form of C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were within normal range.

The patient was treated with anti-coagulation with a plan to maintain the international normalized ratio (INR) in the therapeutic range of 2.5 - 3.5.

Because the patient had severely compromised lung in the presence of extensive bilateral destruction of both lung fields, a further insult of the lung in the form of pulmonary embolism could have been fatal; hence, an inferior vena cava (IVC) filter was implanted to prevent further lung compromise. The IVC filter was planned to be kept in situ at least until the time the varicose veins were not completely treated and no recurrence of embolic events was documented for a significantly long period (Figure 3).

The patient was discharged on oral anticoagulation and has been maintaining stable condition as well as apparently asymptomatic at 12 months of follow-up with no further episodes of embolism. The pulmonary parenchymal disease appeared to be chronic in nature and so the patient was planned for supportive management and close follow-up.

**DISCUSSION**

There have anecdotal reports of pulmonary embolism associated with KTS but pulmonary parenchymal disease has never been reported so far.³ The presence of an asymmetrical distribution of the emphysematous changes in the lungs, which is similar to the general pattern of distribution of this disease, makes it likely to be correlated to the disease pathology in some way; although it might just be a co-incidence, especially because the lung involvement was more on the opposite side as was the distribution of the cutaneous
manifestations. The pathophysiology of this disease is also less studied upon and systemic involvement of this disease has rarely been reported, which makes the description of this case unique. It is more commonly seen in males with the average incidence in the general population ranging between 20 - 50 per million individuals. Capillary malformations of the port-wine stain type are often associated.

In a study involving 252 patients performed at the Mayo Clinic, port-wine stain was reported to be present in 98% of patients; while in another study, which involved 144 patients of KTS, 95% had a cutaneous vascular malformation. In this case, surprisingly there was no port-wine stain which makes it an extremely rare variant.

Involvement of the lower extremities is more frequent as compared to the upper extremities. The patients have small angiokeratomas which may be prone for prolonged bleeding following trauma which forms another concern and sometimes life-threatening complication of the disease.

The appearance of varicosities which may be complicated by pain, thrombophlebitis, and hemorrhage may be delayed up to late in childhood or adolescence. The distribution of varicosities may be asymmetrical and is predominantly seen to involve the lower extremities.

There have been reports in which the presence of other co-existent anomalies such as blue naevi, pulmonary vein varicosities, cerebral aneurysm, gastrointestinal haemorrhage, has been described. Systemic involvement and involvement of lungs has not been previously discussed.

Complications, which can be associated with this syndrome, are thrombophlebitis, dermatitis related to stasis of venous blood, cutaneous ulcers, bleeding, coagulopathy, and congestive heart failure.

Regular clinical and radiographic monitoring by means of duplex scanning contrast venography, ultrasonography, contrast venography, arteriography, and nuclear magnetic resonance imaging (MRI) studies, is essential of which the non-invasive modalities are often preferred. The plasma d-dimer levels can be used to screen the tendency for venous thromboembolism.

Treatment of this disease is essentially conservative. Compression stockings and intermittent pneumatics are useful in cases with limb hypertrophy varicosities. The role of surgery in the management of varicosities and limb hypertrophy is controversial although some have reported use of laser guided surgical therapy for varicosities, which was also tried in our case.

Pulmonary embolism requires chronic anti-coagulation while the role of inferior vena cava filter is controversial although it may be tried in cases where there are contraindications for anti-coagulation or in cases where the pulmonary parenchyma is so compromised that another episode of embolism might be life threatening, as was done in our case.

The management of interstitial lung disease is controversial and depends on the type of involvement and the signs of systemic inflammation. This case was managed conservatively as the pulmonary parenchyma was extensively destroyed and there was no evidence of systemic inflammation which proved the chronicity of the illness.

The co-existence of this syndrome with interstitial disease has not been described previously so it deserves mention although further case reports might add weight to the pathophysiology involved in the co-existence of these diseases, and to confirm whether the diseases were actually correlated from a pathophysiological point of view or the occurrence was just coincidental.

REFERENCES