

KLIPPEL - TRENAUNAY SYNDEROME

A CASE REPORT

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In their basic work, "Du Naevus Variquex Osteo Hypertrophique", published in 1900 in ' Archives Generales de Medicine' French physicians Klippel and Trenaunay described a clinical syndrome with three major symptoms: 1-hemangiomas, 2-hypertrophy of the soft tissue and bone with overgrowth of the extremity, and 3- varicose veins. This syndrome is often mentioned together with the " hemangiectatic hypertrophy" described by Parkes Weber in 1907 and 1918⁵. However, the differences in hemodynamic pathology, prognosis, and treatment of these two diseases seem to be significant. In their original paper, Klippel and Trenaunay do not mention any sign of arterial dilatation, bruit, or localized murmur of pulsating veins, although the clinical picture of arteriovenous (A - V) fistula was already well known at the turn of the century. To contrary, Parkes Weber called the syndrome of hemangiectatic hypertrophy a congenital " phlebarterectasy, " which is dilatation of arterial and venous trunks... with tumor-like hemangiomatous overgrowth in vascular system. " He clearly describes the signs of arteriovenous fistula, in which " sometimes... a definite kind of thrill or pulsa-

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tion... is transmitted to the veins."

Several classifications of the congenital vascular anomalies were published in the literature, but the confusion about the two syndromes still persists. The terms congenital angiectatic hypertrophy,⁹ infatible angiectatic osteohyperplastic syndrome,³ osteoangiohypertrophic syndrome, and Klippel-Trenaunay-Weber syndrome,⁴ have all been used. However, it is widely accepted that the existence of functional arteriovenous shunts determines not only the treatment, but the outcome of the disease.

Therefore this paper reserves the term Klippel-Trenaunay Syndrome, or osteoangiohypertrophy-type "Klippel-Trenaunay," for the limb hypertrophy and varicosity associated with hemangiomas. The historic, hemodynamic, therapeutic, and prognostic differences between the Klippel-Trenaunay and the Parkes Weber syndromes are emphasized by several authors.¹⁴⁻¹⁵

In recent years the development of isotopic diagnostic procedures allowed detection of "hypoactive" or "inactive" arteriovenous fistulae, in some cases of Klippel-Trenaunay syndrome.⁵ However the shunt volume is significantly lower than that observed in Parkes Weber syndrome, and inactive A-V fistula has no hemodynamic effect and no surgical or prognostic consequences.

Case Report:

In 1982 a 17 years old boy (A.A.) was admitted to Valiasr General Hospital, with rectal bleeding and anemia.

He had multiple portwine flat cutaneous hemangioma on the right side of his body (fig 1) which had appeared at the age of six.

There was no history of birth trauma, and apparently no family history.

There was significant varicosity in his right lower extremity. Edema was present in this extremity. The length of right lower extremity was 1/5 Cm greater than the left. (Fig.2-3)

Proctoscopic examination revealed internal hemorrhoids as the cause of rectal bleeding.

The additional change was mild strabism and atrophy of his left testicul without varicocele.

The laboratory tests were normal, including (T3, T4, FSH, LH, Prolactin, Testosterone, and Cortisol,) but hemo globin was 9.0 gr per 100ml and hematocrit was 28 per cent.

Radiographs of chest, hands, skull, were normal, but in left tibia displastic fibrosis (fibroxanthoma) was noticed. (Fig.4)

Superior mesentric angiography and right femoral arteriography were performed, no A-V fistula was found. (fig. 5)

Venography of right lower extremity was performed, which showed deep venous insufficiency with incompetent perforators. (fig 6-7).

The patient was treated nonoperatively, he was advised to wear elastic support, and to use a shoe with higher length on the left foot, iron and diuretic. He is now in good general condition, and his hemoglobin is 11 gr per 100 ml.

Discussion :

The etiology of the Klippel- Trenaunay syndrome is

unknown. No hereditary factors could be confirmed and the exact time of the embryonal maldevelopment is also questionable. The hemangiomas of the different angiodyplasia are never real tumors, but only the result of faulty autogenesis.¹¹ The differences in vascular anomalies are due to the stage of development when aberration from the normal occurred. An excellent review of the etiologic and morphogenetic hypothesis was published by Malan and Puglionisi.¹¹ Vollmar¹⁵ states that an "inborn error" of tissue composition and distribution results in vascular anomalies and skeletal overgrowth, and the latter is not a secondary phenomenon of the vascular disorder, as it is in the case of an arteriovenous fistula.⁷ His hypothesis is supported by the fact that, in Klippel-Trenaunay syndrome, the intraosseous involvement by hemangiomas is rare. There is usually no bone anomaly, except its hypertrophy in diameter and length. In Parkes Weber syndrome, the existence of intraosseous A-V hemangiomas, especially next to the epiphyseal plate, is frequent. Horton, who already in 1934 had reported on 38 cases of congenital A-V fistulae, emphasized that an increased blood flow to the epiphyseal line causes abnormal growth of bone.⁵ Also, from the Mayo Clinic, Janes and Musgrove in 1949 succeeded in creating overgrowth of the hind leg of young dogs by artificial A-V fistulae.² Hutchison and Burdeax were able to produce overgrowth of the hind limb in dogs when they created venous stasis by ligating veins or applying a tourniquet. Nevertheless, it still has not been proven whether the increased skeletal growth

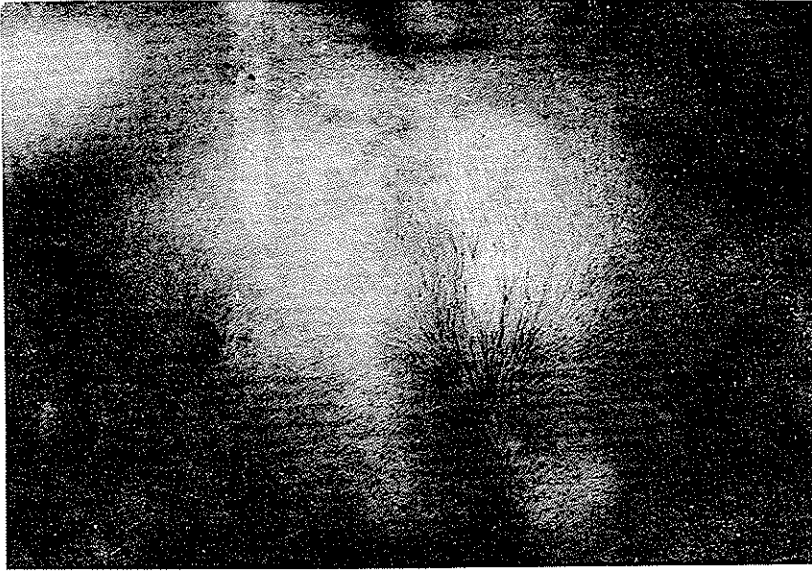


Fig. 1.



Fig. 2

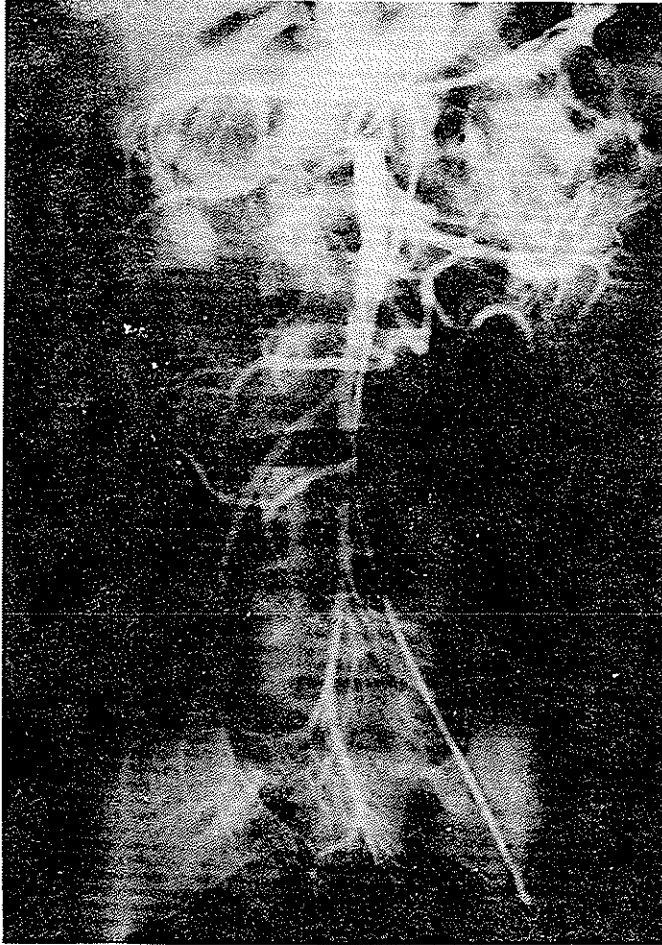


Fig. 5

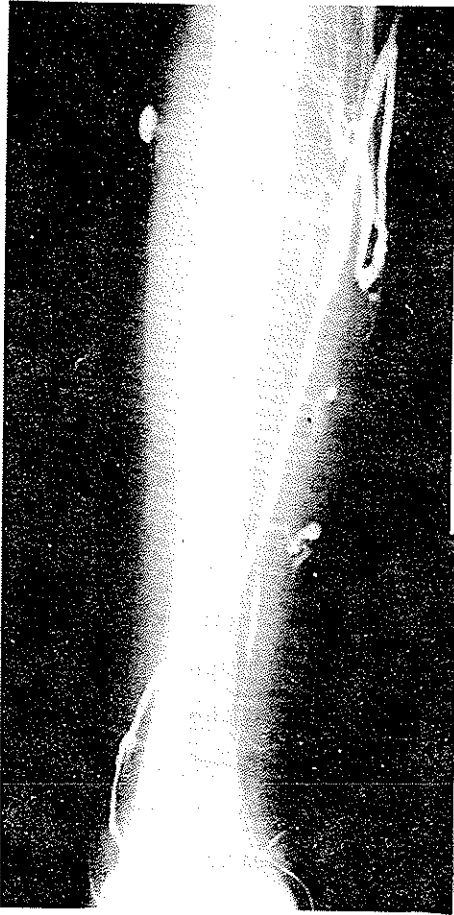


Fig. 6

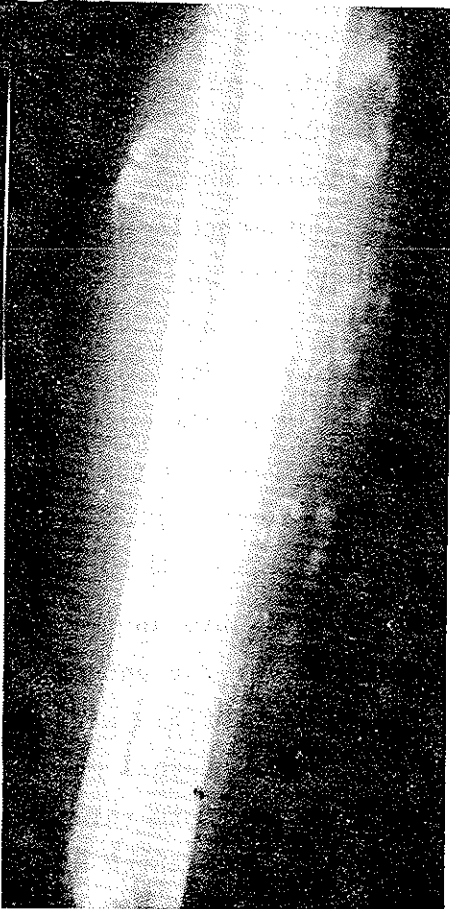


Fig. 7

is caused by an increased arterial flow in the area of the epiphyseal plate, by the venous hyperemia and stasis caused by A-V fistulá, or by an inborn error in tissue development.

To establish the diagnosis of Klippel-Trenaunay syndrome, the presence of the three main symptoms-- varicosity, hemangioma, and extremity hypertrophy-- are necessary. Involvement is usually unilateral, but the rare bilaterl or simultaneous upper and lower extremity involvement have also been mentioned.

The varicosity is usually atypical, often being significant on the lateral aspect of the extremity, with the socalled "lumbar-to-foot" pattern. Dodd and Cockett² call this synrome the "lateral venous anomaly", to emphasize the persistence of the embryonic dorsal or sciatic vein system that normally disappears in the second month of intrauterine life. The persistance of lateral or primary marginal veins can be found in upper and lower extremity deep vein hypoplsias or aplesias in Klippel-Trenaunay syndrome.

The most common form of hemangioma is the capillary type, or port wine nevus, which has a pink-to-purplish color represents diffuse telangiectasias of the superficial vessels of the dermis. The flat purple patch blanches on pressure and sometimes fades, but usually becomes darker as the patients gets older. Cavernous hemangioma or lymphangioma can also found. The hemangiomatous involvement of the internal organs can lead to severe complications in some very rare cases.

Besides the three main symptoms, additional common features are hyperhydrosis, skin atrophy, verrucae, derma-

titis, thrombophlebitis, cellulittis, and testicular atrophy. In Klippel-Trenaunay syndrome, edema of extremity is often present. Additional congenital anomalies such as syndactylia, spinabifida, and equiovaus have been observed in some cases.

Rectal bleeding and hematuria are rare, but are serious complications of the Klippel-Trenaunay syndrome.⁵ In some cases occur due to pelvic or abdominal hemangioma. This can also occur because the dysplastic posterolateral veins of the extremity drain the blood to the hypogastric vein. With this report 12 patients were observed with rectal bleeding.⁵⁻¹⁴

Among the noninvasive diagnostic procedures, doppler flow measurements and venous occlusion plethysmography are useful in the exclusion of A-V fistulae. Quantitative values of arteriovenous shunt volume can be given by isotope diagnostic methods, using radioactive-labeled small microspheres.¹¹ Arteriography is particularly useful in demonstrating large caliber A-V fistula and thus is important in separating Klippel-Trenaunay from Parkes Weber syndrome, If vein stripping is to be undertaken, venography should also be done to extirpative surgery.

If varicosity is minimal, the hemangioma is a port wine superficial nevus, and the hypertrophy of extremity is under 1cm, the patient usually does well without treatment. If the varicosity is more significant, elastic support should be suggested to the patient. If leg length discrepancy is more than 1.5cm, a lift placed in the shoe on the healthy foot can help to avoid the compensatory scoliosis of the vertebral column. The concomitant skin changes and the rare cases of leg ulcer might also

need local care. Drug therapy should be restricted to treatment of inflammatory changes, cellulitis or anemia due to recurrent bleeding from the angiomatous tissue. Patient who develop recurrent cellulitis should be placed on long-term, prophylactic antibiotic therapy. Steroids and irradiation will not stop the development of visceral hemangiomas, and their use in the treatment of this disease is not indicated.

Varicectomy and vein stripping are usually not indicated in the treatment of Klippel-Trenaunay indicated in the treatment of Klippel - Trenaunay syndrome, unless specific symptom are present. Lindenauer reported on 12 patients who are operated on for varicosity, undergoing multiple ligations and stripping. Eleven of these patients had an increase in ankle edema and a rapid recurrences of the varicosity associated with leg discomfort. Extensive varicectomy should not be performed.¹⁵ According to Vollmar,¹⁵ in every fifth case of Klippel-Trenaunay syndrome there is aplasia or hypoplasia of the femoral or popliteal veins, and excision of the superficial bypassing veins leads to an increase in venous insufficiency in these patients. However, if physical examination and venography show patent deep veins, local varicectomy or even stripping can be performed after careful evaluation in each case.

Complete excision of hemangiomatous tissue is seldom possible and can lead to scar tissue formation, repeated bleeding, pain and walking impairment. Surgery is usually not recommended in cases of large hemangiomas, even if bleeding is a major problem for the patient. Percutaneous injection of thrombotic matter, oblitative

polymers, or sclerosing agents may be of greater benefit in these patients.⁵

Surgical intervention is indicated in some cases to inhibit extreme overgrowth of the extremity. Although a discrepancy of not more than 2 Cm can easily be corrected by placing a lift in the heel of the shoe on the healthy foot,⁶ when a marked discrepancy is noted epiphysiodesis or tibial osteotomy should be considered.

Therapy for Klippel-Trenaunay syndrome is mostly symptomatic. Elastic compression and, in selected cases, varicectomy are available methods of treatment. The ideal operation in cases of hypoplasia or aplasia of the deep veins would be venous reconstruction, but this is a task for the future. Severe forms of the Klippel-Trenaunay syndrome rarely occur. Fortunately, because of the benign course of this disease, the majority of these patients do not need surgery and do well with conservative therapy

Summary

The Klippel-Trenaunay syndrome, first reported in 1910, is a triad of cutaneous hemangioma, varicose veins, and soft tissue or bony hypertrophy. The varicosities are extensive involving the tributaries of the greater or lesser saphenous systems. Arteriogram are normal with no evidence of arteriovenous fistulas. Multiple ligation and stripping may worsen the condition, and treatment consists of wearing elastic supporting hose. A similar condition but associated with arteriovenous fistula is known as the Parkes Weber syndrome.

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