ORIGINAL RESEARCH PAPER

Volume-7 | Issue-6 | June-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PARKES WEBER SYNDROME WITH SYNDACTYLY- AN UNUSUAL PRESENTATION: A RARE CASE REPORT

Dermatology

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ABSTRACT

Parkes weber syndrome is a rare vascular disorder characterized by arteriovenous malformation and limb hypertrophy with close clinical resemblance to Klippel Trenaunay syndrome. Syndactyly is a feature often reported in cases of Klippel Trenaunay syndrome. We report a case of Parkes weber syndrome in a 7 year old boy with syndactyly of the toes of right foot

KEYWORDS

Syndactyly, Parkes Weber Syndrome, Klippel Trenaunay Syndrome

Introduction:

Parkes weber in 1918, described this condition first as a haemangiectatic hypertrophy where there is limb swelling along with arteriovenous anastomoses. It is differentiated from Klippel Trenaunay syndrome by the presence of A-V malformation instead of a predominantly venous malformation. There is hypertrophy of the affected limb due to the increased localized blood supply. Syndactyly has not been reported in conjunction with Parkes Weber syndrome; here we report a case of Parkes weber syndrome in a 7 yr old boy with syndactyly of toes and hemangioma of the tongue.

Case report:

A 7year old boy was brought to the skin OPD with complaints of fleshy swelling over the dorsum of the tongue present from 3 months of age. It was initially small in size but gradually enlarged to the current size. The lesion was painless with no history of trauma or spontaneous bleeding. There was no history of similar complaints in other family members and patient was born out of non - consanguineous marriage. On examination a well defined erythematous plaque with well defined border of size 6*4cm present over dorsum of tongue. No visible pulsation. On palpating the plaque was firm. No tenderness or bleeding. Further examination revealed fusion of 2^{nd} 3^{rd} and 4^{th} digits of right foot. Hypertrophy of 3^{rd} digit of right hand. On measuring the upper limb there was increase in girth of right wrist by 2cm when compared to the left. Gait was normal. No varicosities noted. Systemic examination was normal. Skin biopsy was taken from 3^{rd} digit of right hand where histopathology revealed dilated blood vessels in upper and mid-dermis with no proliferation.

Discussion:

Parkes weber syndrome is an autosomal dominant disorder. It is a vascular anomaly which is characterized by arteriovenous fistula, hemangioma and soft tissue as well as bone hypertrophy where the affected limb is larger than the other. Upper limb is most commonly affected than lower limb [3]. It is usually present from birth. It is often confused with Klippel Trenaunay syndrome where there are venous malformations and other similar features except arteriovenous fistula which is the differentiating feature. Syndactyly is usually a feature of Klippel Trenaunay syndrome [1]. Complications of Parkes weber syndrome include congestive cardiac failure, abnormal bleeding, Arteriography and skin biopsy can be done to distinguish from Klippel Trenaunay syndrome. MRI soft tissue can be done to assess the limb discrepancy. Supportive therapy by advising them compression garments. Customized footwear should be advised to correct limb discrepancy. It is medically managed with the aim of decreasing the preload by giving intravenous labetalol 1mg/min using infusion pump. Surgically it is managed collectively by interventional radiologist and vascular surgeons. Embolization is another surgical option where we can combine with surgical resection targeting occlusion.

Differential diagnosis includes Klippel Trenaunay syndrome and Proteus syndrome

CONCLUSION:

This case is presented because of the unusual and rare manifestation syndactyly (fusion of three digits of right foot) along with haemangioma of tongue. It has to be differentiated from Klippel Trenaunay syndrome.

Figure 1: Clinical picture showing:

A) Hypertrophy of the 3^{rd} digit of the right hand
B) Syndactyly noted over right foot (fusion of 2^{nd}, 3^{rd}, $4^{th digits}$)
C) Hemangioma over dorsum of the tongue

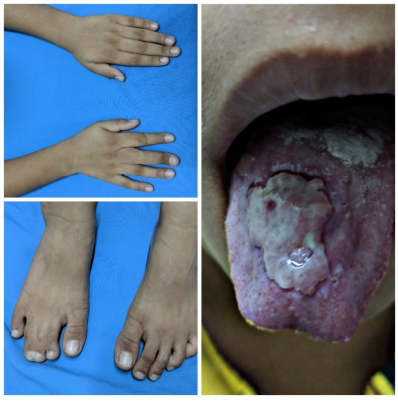
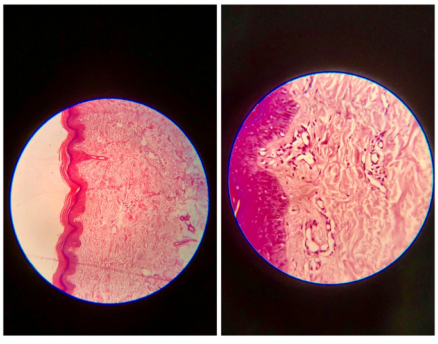


Figure 2: A and B under low power and high power respectively showing multiple dilated blood vessels in upper dermis and mid-dermis.



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