Laparoscopic Management of Klippel Trénaunay syndrome Anorectal haemangioma

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•**Introduction** -Klippel Trénaunay Syndrome is a congenital condition characterised by vascular malformations, venous varicosities with pathological soft tissue growth. **congenital**
•**TRIAD OF 1)mixed vascular malformation(capillary hemangioma , port wine stain) 2) Venous varicosities with 3) abnormal soft tissue growth and or bone growth also called "angio-osteo-hypertrophic syndrome" (AOH) "venous**
• osteohypertrophicus" "hemangectasia hypertrophicans"

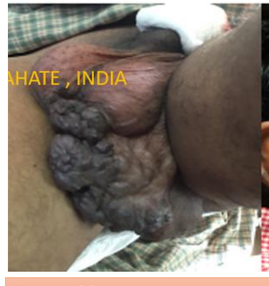
Case

A 50 year old gentleman came with complaints of recurrent rectal bleeds, multiple haemangiomas on left lower limb, scrotum and face. On examination. Bleeding rectal and anal haemangiomas were demonstrated.

Principle

The intention of laparoscopic management is to remove from the lower segment of the rectum; the diseased, dilated, and tortuous vessels; and the adjacent tissues, hypertrophied and consolidated by plastic exudation. These diseased structures are situated beneath the mucous membrane of the rectum, and rest upon the circular fibers of the internal sphincter; and it is the objective of the operation to clear them away from the anal orifice, where they act as obstructions to defecation, and excitants of periodic and distressing spasmodic muscular contractions.

Result



Scrotal hemangioma



facial hemangioma



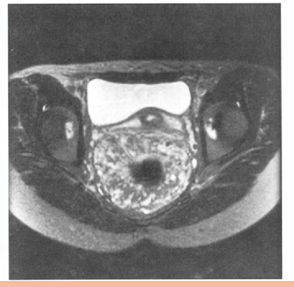
foot Boney deformity



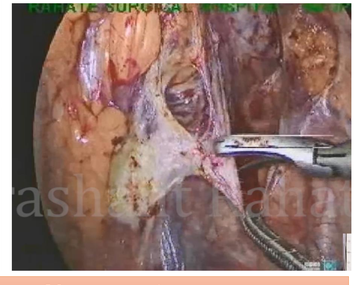
Colonoscopic vision-haemangioma up to sigmoid



MRI rectal haemangioma



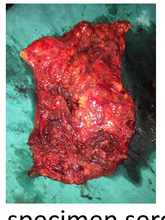
laparoscopic view serosal extension of haemangioma







specimen
mucosal side



specimen serosal side

Rectal haemangiomas have often been misdiagnosed as internal haemorrhoids, inflammatory bowel disease, portal hypertension, carcinoma, and other conditions angiomas have often been misdiagnosed as internal haemorrhoids, Laparoscopic approach for excision up to the extension of haemangioma in bowel .During laparoscopy hormonic scalpel is used with 1 & 3 mode – haemostatic mode compression by gauzpiece helped haemostasis. Laparoscopic APR resulted in complete alleviation of symptoms like recurrent rectal bleed and alarming anaemia , recurrent hospitalisation and recurrent blood transfusions