On-line Table: Reported cases of KTS and spinal AVMs

Report No.	No. of Patients	Age, Sex	SAVM Type and Location	Authors' Presumed Signs of KTS	Signs Incompatible with KTS	Possible Alternative Diagnosis[a]
1) Rohany et al[4]	2	Case I: 37-Y-F	Case I: T12-L3 single-hole spinal cord fistulas	Case I: limb hypertrophy, surgical removal of "venous malformations"	Case I: venous varices limited to foot, foot was warmer, no evidence of LM or CM	Case I: NA
		Case II: 29-Y-F	Case II: C6-T1 AVM drained to paraspinal veins and fed by branches of vertebral and costocervical arteries	Case II: multiple VMs of extremities, chest wall, spleen, and pulmonary parenchyma with pulmonary vein varicosities	Case II: All 4 extremities and trunk were involved, pulmonary lesions and varicosities, no VM or CM	Case II: NA
2) Alexander et al[5]	1	30-Y-M	T3-T4 paraspinal AVM	VMs, cutaneous "hemangiomas," and hemihypertrophy	Unusual bilateral syndactyly and macrodactyly with port-wine stain of trunk	CLOVES syndrome (paravertebral lipomatous mass insinuating into spinal canal, scoliosis, furrowed soles, bilateral foot/limb anomalies, paraspinal AVMs)
3) Brunaud et al[6]	1	39-Y-M	Thrombosis of anterior spinal artery/artery of Adamkiewicz	Arm hypertrophy with disseminated cutaneous "angioma"	Cerebral ischemia from internal carotid dissection, hemihypertrophy, bilateral leg hypertrophy, and varices; lack of LM or CM	NA
4) Nakstad et al[7]	1	13-Y-F	T11-L2 dural fistulas	Right leg "gigantism," skin angiomas of tface and gluteal region	Facial birthmark, blindness, and increasingly progressive overgrowth	NA
5) Kojima et al[8]	1	28-Y-F	T11-L2 posterior extramedullary AVM	Nevus flammeus, varices, and hypertrophy of leg	Warmer leg and atypical distribution of cutaneous stain	CM-AVM (*RASA1* mutation)?
6) Szajner et al[9]	1	48-Y-F	Several AVFs supplied by thyrocervical and costocervical trunks	Cutaneous and subcutaneous "angiodysplasia" of left hemithorax	Fatty overgrowth of paraspinal region with nonspecific "angiodysplasia"	NA
7) Arai et al[10]	1	37-Y-M	"Arteriovenous hemangioma"	Truncal tumor, "angiomatous nevi," right leg hypertrophy, and deformities	Huge infiltrative truncal mass insinuating into spinal canal with small truncal stain	CLOVES syndrome (paravertebral lipomatous mass insinuating into spinal canal, severe scoliosis, foot/limb anomalies, paraspinal AVMs)
8) Benhaiem-Sigaux et al[11]	1	9-Y-F	T11–12 intramedullary AVM	Hypertrophic right leg, varix, and cutaneous angioma of leg and flank	Diagnosis is vague, no photos or proof of embryonal veins or LM	NA
9) Djindjian et al[12]	5	Case I: 13-Y-M	Case I: T9-T12 spinal AVM	Case I: superficial varicosities	Case I: arteriovenous fistula of leg, vertebral "hemangioma," lack of LM or CM	Case I: NA
		Case II: 19-Y-F	Case II: T4+5 spinal AVM	Case II: right arm phlebectasia and extensive AVMs	Case II: extensive AVM of arm, subclavian ectasia, subarachnoid hemorrhage, vertebral erosions and limb; photo and bony changes on arm; x-rays are not those of KTS	Case II: NA
		Case III: 19-Y-M	Case III: T9-L2 intramedullary "angioma"	Case III: cutaneous angioma of thigh and umbilicus	Case III: distribution of "angioma," warmer limb, marked kyphoscoliosis, and subarachnoid hemorrhage	Case III: NA
		Case IV: 15-Y-F	Case IV: L1–2 intramedullary AVM	Case IV: L1–2 intramedullary AVM	Case IV: subarachnoid hemorrhage, knee AVM, family history of premature death from intracranial hemorrhage are not KTS features; photo is suggestive of ischemic ulcerations; lack of classic CM and LM	Case IV: NA
		Case V: 15-Y-M	Case V: lower spinal cord and cauda equina AVM	Case V: hypertrophic right leg with varices	Case V: numerous AVMs of leg	Case V: NA
10) Rodesch et al[13]	3	NA	NA	No data presented to support diagnosis of KTS	No data given	NA
11) Schulz and Neumann[14]	1	24-Y-M	T6–8 vascular malformation-necrotizing myelopathy (Foix-Alajouanine syndrome)	Arteriovenous shunts, varicose-like superficial venous system, warmer leg	Increased flow velocity in leg arteries/arteriovenous shunts; warmer leg and generalized sweating are not signs of KTS; photo, poor quality, not typical	CM-AVM (*RASA1* mutation)?
12) Forster and Kazner[15]	1	10-Y-F	T11-L2 spinal angioma	Larger right leg with nevus flammeus, scarred skin with "erosions"; dorsum of foot and toes were edematous	Pattern of overgrowth in photos, edema/swelling of dorsum of foot and toes, impalpable foot pulses not typical signs of KTS	CM-AVM (RASA1 mutation)?

Table continues

On-line Appendix

On-line Table: Reported cases of KTS and spinal AVMs *(Continued)*

Report No.	No. of Patients	Age, Sex	SAVM Type and Location	Authors' Presumed Signs of KTS	Signs Incompatible with KTS	Possible Alternative Diagnosis[a]
13) Gourie-Devi and Prakash[16]	1	25-Y-M	C7-T2 epidural "hemangioma"	Deep purple nevus of shoulder and upper extremity	Birthmark, distribution, and lack of VM and CM are not typical of KTS	NA
14) Carter et al[17]	1	20-Y-M	T5-L2 hypervascular tumor	Right leg hypertrophied, venous varicosities and supernumerary toes	Bilateral thoracic cutaneous "hemangioma" and angiolipomas are not typical for KTS	CLOVES syndrome?
15) Pichierri et al[18]	1	NA	C5–6 intramedullary cervical cavernoma	Right bony and soft-tissue hemihypertrophy and cutaneous angiomatosis of right hand; cutaneous telangiectasias of right leg	Right arm hemihypertrophy and cutaneous "angiomatosis" and cutaneous telangiectasias are not KTS features	NA
16) Tan et al[19]	1	19-Y-M	T2–6 juvenile-type spinal AVM	Right lower limb progressively enlarged	Multiple warts of face, trunk, and right extremities and warmer leg are not KTS signs	NA
17) Rosenblum et al[20]	1	NA	Intradural SAVM	NA	No data given	NA
18) Den Hartog Jager[21]	1	27-Y-M	T9–10 "intradural angioma venosum"	Congenital "lymphangioma"; asymmetric legs and telangiectasia	Lack of classic venous anomalies and lymphatic malformation; photos are not supportive of KTS	NA
19) Eber et al[22]	1	32-Y-M	AVM of conus medullaris	Left lower extremity "angioma" and varices	Facial "angiomas" and varices, pelvic AV fistulae, claudication, and hyperdynamic circulation are not features of KTS	NA
20) Pitagoras de Mattos[23]	1	33-Y-M	T9–10 unspecified "diffuse angioma"	Red discoloration of skin, several red irregular lesions ranging from 5 to 20 mm in face, back, arms, and legs; atrophy of lower extremity and scoliosis	Progressive nature, multiple small stains in hemifacial and periauricular distribution; similar lesions on back, arms, and legs; atrophy of lower extremity and scoliosis are very unusual presentation of KTS	CM-AVM?
21) Vajda and Brozmanova[24]	1	10-Y-M	T12-L2 "hemangioma"	Right leg overgrowth and vascular stain	Lumbosacral geographic "hemangioma" extending into medial side of right lower extremity; warmer affected leg and tethered cord are not features of KTS	CLOVES syndrome?
22) Jyoichi et al[25]	1	67-Y-M	Small AVM supplied by artery of Adamkiewicz	Vascular birthmark and varicose veins of left leg	AVM of left iliac vessels, stroke, hearing loss, lack of lymphatic component, and cyanosis of limb are not features of KTS	CM-AVM?
23) Fukutake et al[26]	1	47-Y-M	Left C5 and T4 nidus-like blush	Port-wine stain of right leg and upper back with venous dilation	Combined upper and lower limb syndactyly, preferential overgrowth below knee, lack of LM, and faint color of port-wine stain are not features of KTS	NA
24) Sharma[27]	1	16-Y-M	Multiple intradural spinal cord AVFs and renal artery aneurysms	Progressive weakness of bilateral lower limbs with marginal vein	Nevoid geographic skin lesion shown in photos is not classic for KTS, no definite overgrowth of fatty and lymphatic tissue, presence of marginal vein is not pathognomonic of KTS	NA

[a] ? indicates "not certain."

On-line Appendix

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