Reference

1. Leach RD, Eykyn SJ, Phillips I, Corrin B, Taylor E A. Anaerobic axillary abscess. Br Med J 1979; 2: 5-7

KLIPPEL - TRENAUNAY SYNDROME

To the Editor,

A one-year-old boy a product of consanguinous marriage, presented with an abnormally large left foot since birth. There was no positive family history of similar disease. There was grotesque enlargement of the left lower limb with increased length and girth. Hypertrophy of the left foot with macrodactyly and normal movements at the subtalar and ankle joints were seen. Single port-wine stain with irregular margins was present over the right lumbar region. There was no evidence of varicose veins, abnormal pulsations or bruit over the left lower limb.

Routine haemogram, urinalysis, liver enzymes estimation, serum calcium and phosphate levels and ultrasonography of abdomen were normal. There was nothing abnormal in X-rays of the skull and chest, while X-ray of left foot showed evidence of soft tissue overgrowth and increased transverse diameter of phalanges suggestive of macrolipomatosis dystrophy.

The association of varicose veins, soft tissue and bony hypertrophy, and cutaneous haemangioma of the port-wine variety confined to one extremity was first reported in 1900 by Klippel and Trenaunay.[1] However, not all patients have all the abnormalities of the triad.[2]

The interesting and unusual features of this rare case were the coincidental history of consanguinity in the parents, presence of a single port-wine stain on the contralateral side of the body away from the affected limb, absence of varicosity and bruit over the left lower limb, and associated macrodactyly, in addition to the bony and soft tissue hypertrophy of the left lower limb.

K Krishna
Pune

References

1. Lindenauer SM : The Klippel-Trenaunay-Weber Syndrome: Varicosity, hypertrophy and haemangioma with no AV fistula. Ann Surg 1965; 162 : 303-14.
2. Koh HK, Bhagwan J : Tumours of the skin. In: Dermatology (Moschella SL and Hurlay HJ, eds). 3rd edn. Philadelphia: W B Saunders Company, 1992; Vol 2: 1721-1808.

BLEPHAROCHALASIS

To the Editor,

A 25-year-old man gave a history of repeated transient attacks of painless swelling, of eyelids of both eyes since the age of 15 years. Such episodes progressively become more frequent with age. There was no history of similar disease in family members, or any preceding emotional or physically traumatic event. There was no evidence of eyelid trauma, infection, contact eczema, angioneurotic oedema, cutis laxa or lip swelling. Investigations for evidence of tracheobronchomegaly, goitre and renal disease were negative. Bilaterally symmetrical lid laxity, predominantly of the lower eyelids, with thinning, atrophy, wrinkling and prolapse of the orbital fat was present giving the patient an appearance of tiredness and premature aging.

Blepharochalasis is laxity of the eyelid skin due to a defect in the elastic tissue. It occurs in young people around puberty.[1] Its cause is unknown. Most cases are sporadic, but some pedigrees show autosomal dominant inheritance.[2] Many develop blepharochalasis after an emotionally or physically traumatic

event.[3]

Lower eyelid are affected only in severe cases, and unilateral disease is uncommon.[3] This rare case of blepharochalasis was unique in that the lower eyelids were involved predominantly with lesser involvement of the upper eyelids, and there was no associated pigmentation or telangiectasia.

K Krishna
Pune

References

1. Brazin SA, Stern LJ, Johnson WT. Unilateral blepharochalasis Arch Dermatol 1979; 115: 479-81.
2. Burton JL, Ebling FJG. Disorders of connective tissue. In : Textbook of Dermatology (Rook A, Wilkinson DS, Ebling FJG, et al, eds), 4th edn. Oxford : Blackwell Scientific Publications, 1986; 1787-858.
3. Custer PL, Tenzel RR, Kowalczyk AP. Blepharochalasis syndome. Am J Ophthalmol 1985; 99 : 424 - 8.

VOGT-KOYANAGI-HARADA SYNDROME

To the Editor,

Vogt-Koyanagi syndrome presents as acute anterior uveitis, alopecia, vitiligo, poliosis and dysacousis.[1] Harada's disease presents as posterior uveiltis, meningeal irritation and an increase in concentration of protein and white blood cells in the CSF. The two disorders are found together, hence the term Vogt-Koyanagi-Harada (VKH) syndrome.[2] Meningeal or ocular symptoms usually appear first but in some patients vitilligo may appear before uveitis.[2] It is a rare disease and early diagnosis is important because ocular morbidity can be reduced significantly in some patients with early treatment.[3]

One 35-year-old woman presented with bilateral symmetrical periorbital vitiligo for 6 months and right facial palsy, deafness of right ear, diminished hearing (left ear) and bilateral loss of vision for 2 years and 9 months. Fundus revealed clear media and retina showed resolved oedema with pigment dispersion and few spots of healed juxta-papillary choroiditis.

In February 1992, she was admitted with sudden onset of severe headache, vomiting, moderate grade fever, generalised seizures, loss of memory, speech disturbences, attacks of loss of consciousness with incontinence of urine and stools. At that time CSF had proteins 30mg% suger 73 mg%, chlorides 132 meq/l, 5 WBCs/HPF and no AFB, Gram negative diplococci or cryptococi etc. ELISA for tuberculosis was negative. After 1 month of admission she developed blurring of vision and diplopia which progressed in a month to total loss of vision, first in left eye and then in right. She also had alopecia totalis and hair regrew after 6 months. In April 1992, she had anosmia, loss of taste sensation in anterior 1/2 tongue, nasal regurgitation, complete deafness of right ear, sensorineural deafness of left ear, right 5th, 7th, 8th, 9th, 10th and left 6th cranial nerves palsies with loss of lacrimation. She had dilated 3 mm pupils, diminished light reflexes, Marcus Gunn pupil, bilateral papilloedena, macular oedema, engorged dilated vessels and bilateral upgaze palsy. NMR revealed Arnold Chiari malformations type - I. CT scan was normal. Initially she was treated with 45 mg prednisolone and 300 mg of dilantin daily. Seizures continued and now her seizures are controlled with 300 mg of carbamazepine daily for 1 year. Slight improvement in vision has occurred during the last months. Multiple cranial nerves plasies observed in this patient were unusual. Meningeal disease was severe in our patient.

RR Mittal, JS Jassal, MS Dhaliwal,
GS Randhawa, AK Cropra
Patiala