A case of Radiologically Isolated Syndrome suggestive of demyelinating disease in Klippel-Trenaunay syndrome.

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BACKGROUND

Klippel-Trenaunay syndrome (KTS) is a rare condition characterized by the following triad port-wine stain, varicose vein, and hypertrophy of bones and soft tissues (Fig. 1). We present the first case of KTS showing multiple brain white matter lesions (WML) suggestive of demyelinating disease (MS)).

CASE DESCRIPTION

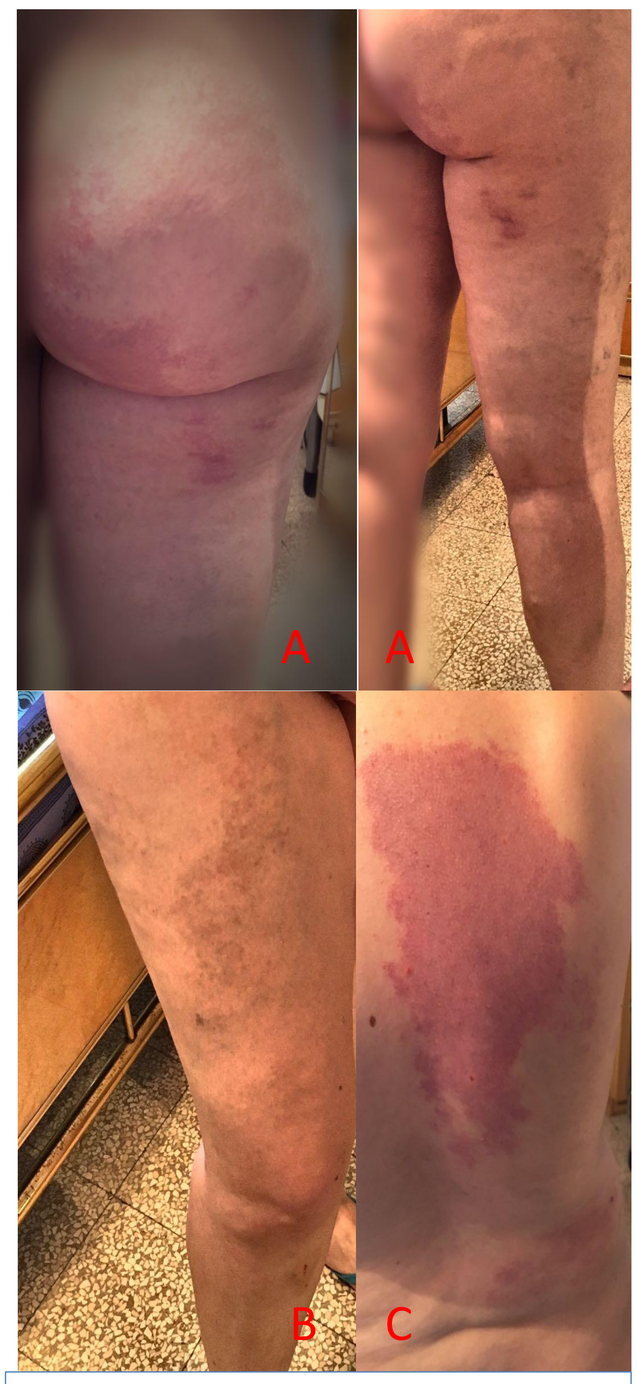

Fig. 1 A cutaneous haemangioma of the gluteus and of the right leg; B varicous veins; C cutaneous haemangioma of the back.

We examined a 33 years old woman with clinically confirmed diagnosis of KTS showing arteriovenous fistulae, asymmetric length and functional impairment of the right leg, many cutaneous haemangioma, varicose veins, episodes of thrombophlebitis and spontaneous fractures in ankle joint. She had a family history of MS (sister and I grade cousin), and was affected by migraine. A brain MRI performed to evaluate the possible presence of vascular malformation, incidentally showed multiple unenhancing WML in centrum semiovale, peritrigonal, cortico-subcortical in frontal, parietal and insular regions (Fig. 2). Neurological examination was unremarkable except for brisk deep tendon reflexes. Routine blood tests were all normal, only anti Yo-Ab were weakly positive. A brain MRI performed 3 months later was unchanged. Spinal cord MRI showed unenhancing lesion in D11-D12 (Fig. 3) . SSEP, VEP, BAEP were normal Patient refused to carry out the lumbar puncture. The patient met Okuda criteria for the incidental MRI anomalies suggestive of multiple sclerosis.

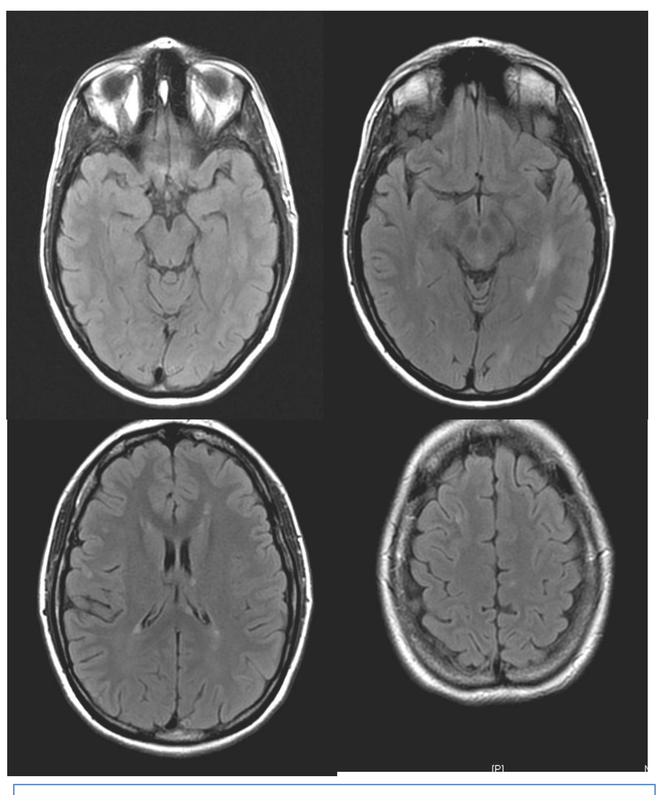

Fig. 2 brain MRI: white matter lesions in FLAIR

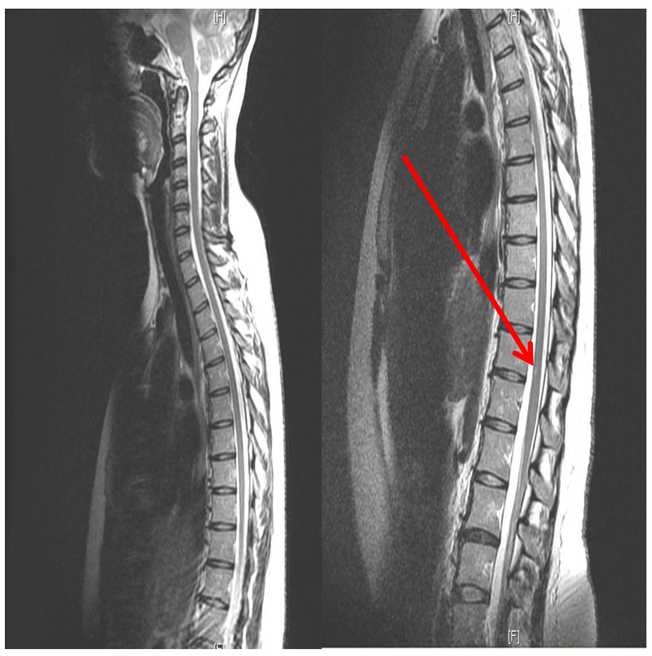

Fig. 3 spinal cord MRI : lesion in D11-D12

CONCLUSIONS

To our best knowledge, this is the first description of radiologically isolated syndrome suggestive of demyelinating disease in a KTS with family history of multiple sclerosis. This reported association might be probably due to chance, but on the other hand this could be a spectrum of a more complex syndrome.

References

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