Neuroradiology (1993) 35: 561–562

Correspondence

Neuro radiology

We read with great interest the paper written by Nakstad et al. [1] concerning multiple arteriovenous fistulae in the Klippel-Trenaunay-Weber syndrome (KTW). This case report raises several questions we would like to discuss.

As emphasised by the authors, KTW is a rare disease which includes venous dysplasia and gigantism of a limb and cutaneous angiomas. Several papers deal with spinal cord arteriovenous malformations associated with KTW [2–5]. We think that the authors' patient cannot be accepted as a cases of this disease, because of the absence of varices and of the unusual anatomical location of the lesion they describe (dural), not previously described in KTW. Rather than multiple dural arteriovenous fistulae, it seems that the lesion illustrated, (although no selective injection of the feeding vessels is shown), is a pial malformation, perhaps fistulous, fed by several converging radiculopial and radiculomedullary arteries [6]. The post-embolisation result shown in Fig. 1 c reveals occlusion of what seems to be the descending branch of a radiculomedullary artery (lying beneath the pial mater), which cannot a dural vessel. The cure achieved suggests a fistulous pial lesion fed by multiple arteries converging on the same point, and secondarily causing congestion of the perimedullary veins.

Furthermore, dural arteriovenous malformations are not seen in children [7, 8]. They are encountered in adults [9, 10] and are rarely multiple. Multiple arteriovenous malformations of the spinal cord have, to our knowledge, never been reported, even in Weber-Rendu-Osler disease. Malformative congenital vascular malformations of the spinal dura mater do not exist, even in Cobb's syndrome, that affects at the same metameric level the skin, bone and spinal cord, sparing the meninges [7, 11].

The fact that postoperative angiographic follow up was performed not selectively but only with unsubstracted aortography, three months after the endovascular procedure, does not prove complete and permanent closure of the fistula. In general, proximal occlusion by coils and balloons, as reported in this case, does not guarantee the stable occlusion of the lesion that can be offered by distal (intranidal) embolisation with glue.

Finally, this interesting paper is very frustrating from both radiological and clinical point of views, as there is no discussion of the gigantism of the right leg (or the hypoplasia of the left), in the apparent absence of varices. In addition, it would be of interest to find an explanation for the right facial angioma and blind right eye (Sturge-Weber syndrome?), and of the association of a right gluteal "angioma" and right leg gigantism (equivalent of osseous hypertrophy seen in haemolymphangioma?)

References

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Reply

Two good things have resulted from our involvement in the case referred to above. Firstly the fistulae are still closed and the patient is doing very well. Secondly our paper has provoked the interesting comments on spinal arteriovenous malformations by Drs Rodesch and Lasjaunias.

We accept the possibility that the fistulae in our case are pial, not dural. However, we would like to make the following additional comments.

Of course the postoperative angiogram was performed with selective injections as well as with overview angiograms. We simply represented the postembolisation angiograms by means of an overview image (our Fig. 1f).

Prominent members of the interventional neuroradiology community repeatedly state that stable occlusion of an AVM can only be offered with glue. We do not think that this is unequivocally documented; on the contrary, we believe that other embolic agents may also be effective. However, of course we agree with Drs Rodesch and Lasjaunias that the embolic agent must reach the fistula or nidus to be effective.

To understand fully angiograms of spinal arteriovenous malformations and to use the correct terminology may at times be difficult. This is probably reflected by the use of such terms as "seems to be", "suggests", etc., in the letter by Drs Rodesch and Lasjaunias. Young et al. [1] illustrate in an excellent way the difficulties of terminology when discussing the Klippel-Trenaunay and Parkes-Weber syndromes. Like Djindjian et al. [2], in two cases with no cutaneous angiomas, we decided to use the term-Klippel-Trenaunay-Weber syndrome in our case, even though one of the classical elements of the syndrome (varices) was absent.

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