

# HIRATA DISEASE AND INTRACRANIAL VESSELS HYPOPLASIA: A CASE OF STROKE MIMIC IN A YOUNG PATIENT

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## Background

The Insulin autoimmune syndrome (IAS) is characterized by spontaneous hypoglycemia caused by insulin autoantibodies in the absence of exogenous insulin administration. IAS is often comorbid with other autoimmune disorders, mostly Graves disease and Hashimoto thyroiditis. IAS is common in Japan where over 200 cases of IAS-related hypoglycemia have been reported. Sulphydryl compounds contained in some drugs like alpha-lipoic acid (ALA) (Figure 1) are known to initiate the onset of IAS. Possibly interacting with the sulphhydryl bonds of insulin (Figure 2).

## Methods

We report the case of a 45-year-old Italian woman referred to our Department because of the sudden onset of a right motor-sensory syndrome on awakening.

## Results

Her past medical history was relevant for Hashimoto thyroiditis, migraine with visual aura, smoking, and no frank diabetes; 15 days before the event she started a dietary multivitamin supplementation containing ALA. On admission, the NIHSS score was 5, plasma glucose level was 36 mg/dL. After glucose infusion a dramatic improvement of neurological symptoms was observed thus avoiding intravenous thrombolysis. Brain MRI including diffusion weighted imaging (DWI) was unrevealing, while intracranial MR angiography (MRA) showed hypoplasia of the left internal carotid artery, the A1 segment of the anterior cerebral artery (ACA) and the left middle cerebral artery (MCA) (Figure 3). Then insulin autoantibodies were detected and IAS was diagnosed.

## Conclusions

In our patient, the right motor-sensory syndrome, worsened after IAS-induced hypoglycemia, occurring in the presence of hypoplasia of the left intracranial carotid circulation revealed by intracranial MRA. Although rare, IAS may be facilitated by commonly used dietary supplementation, such as ALA. This possibility should be considered in young adults with stroke-like syndromes.

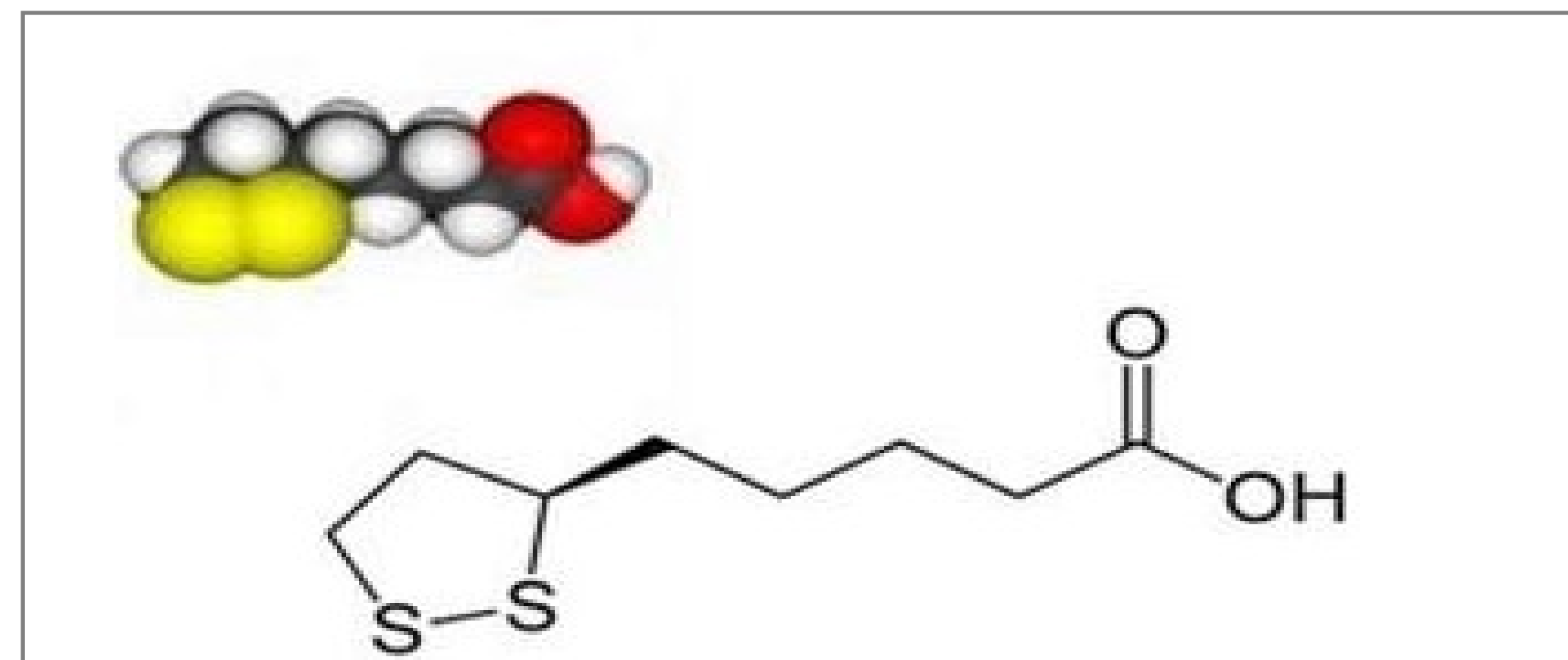


Figure 1

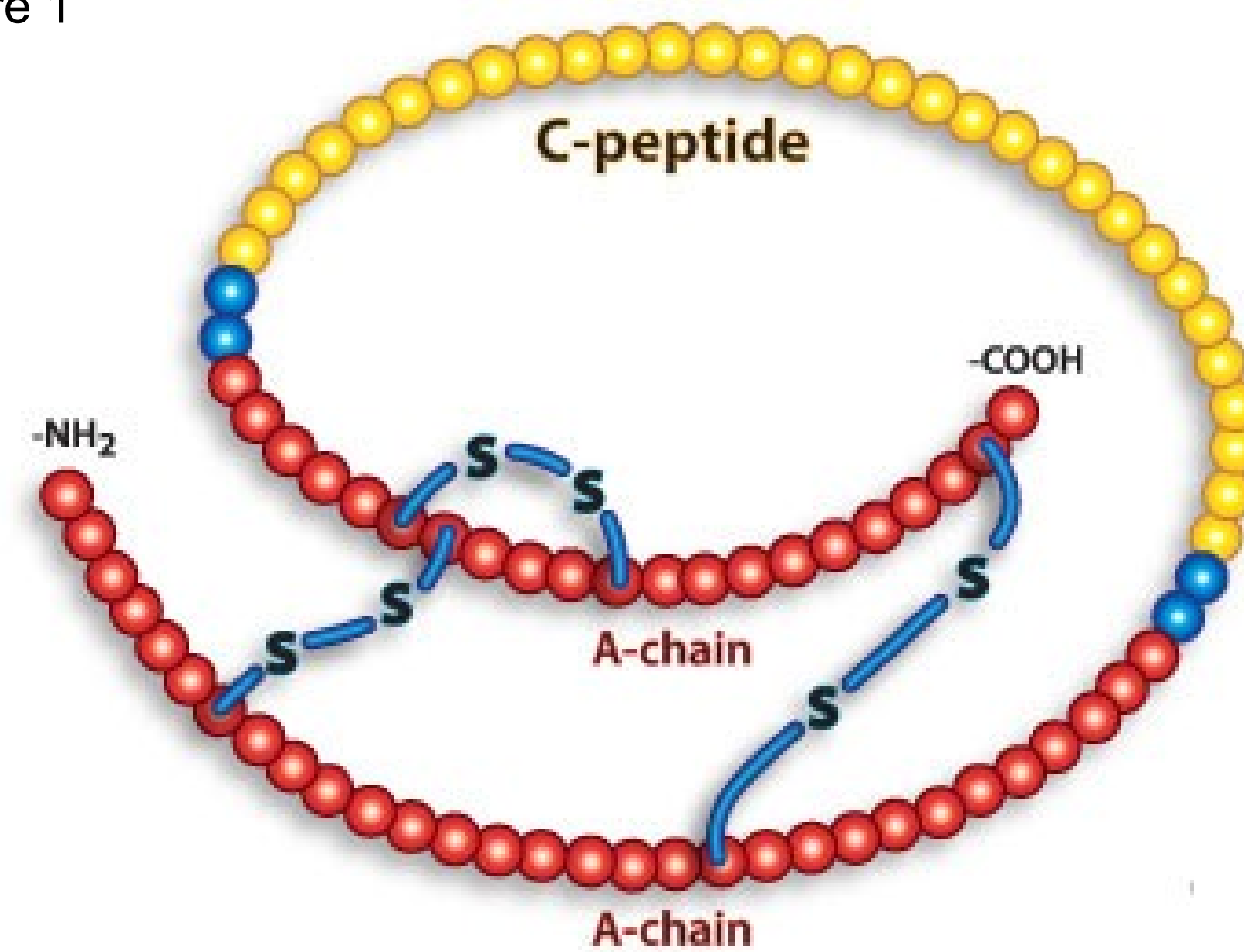


Figure 2

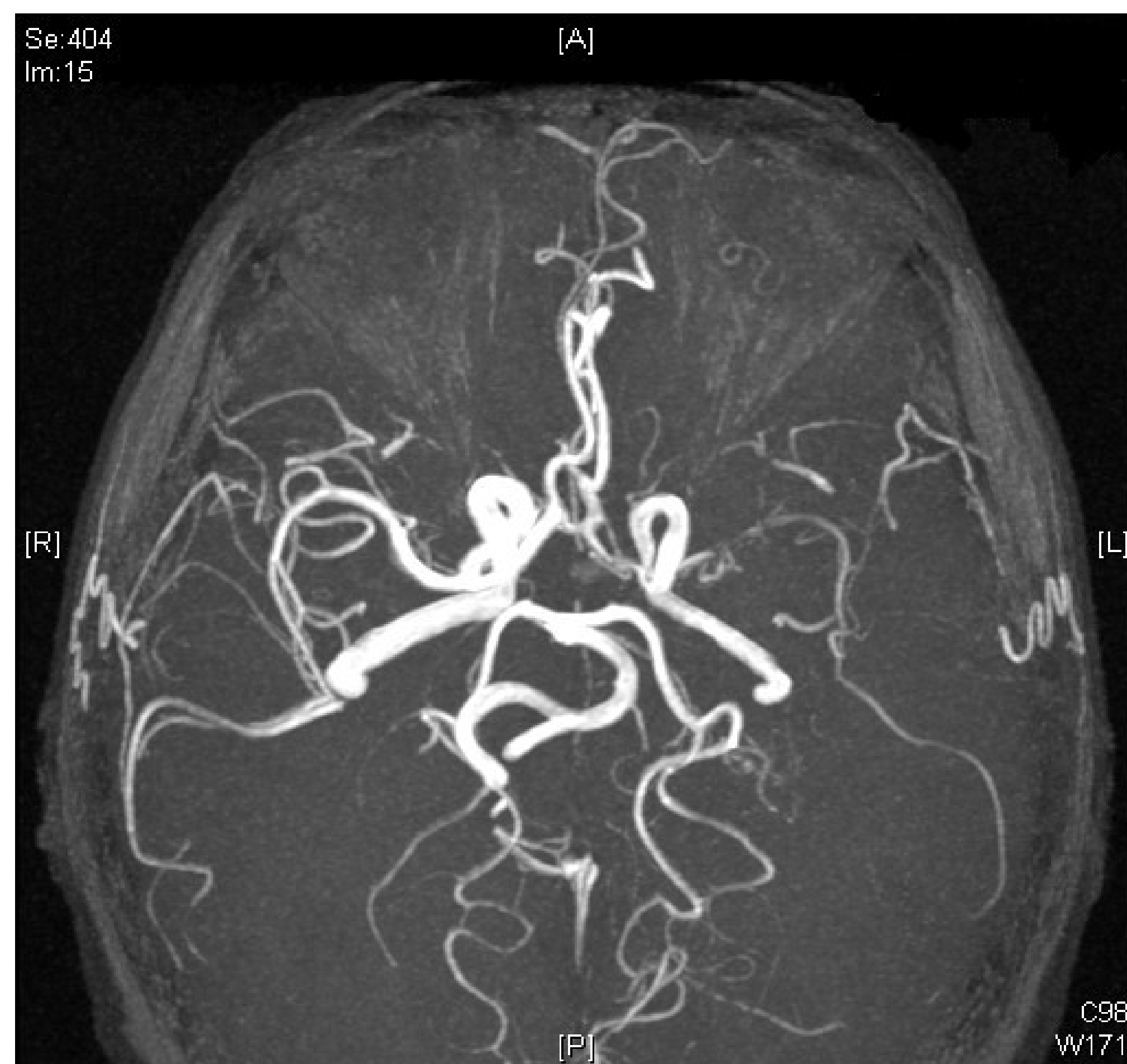


Figure 3

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