

One month after surgery, intravenous immunoglobulin was given biweekly for eight weeks, without clinical improvement, and five months after surgery, seven plasma exchanges were tried, again without clinical improvement. At this point, a second EMG showed severe chronic denervation in C5-C6-C7 territories bilaterally symmetric. The abnormal MRI signal in the left temporal region completely regressed (Figure 1B).

She had a complex partial seizure despite phenytoin. Last EEG showed left subcontinuous slow activity with occasional fronto-centro-temporal sharp waves without PLEDs.

Anti-Hu Ab remained positive seven months after tumor removal.

Mild motor improvement was seen two years after the initial presentation, along with no tumor recurrence.

## DISCUSSION

According to the criteria published by the "Paraneoplastic Neurological Syndrome Euronetwork"<sup>4</sup>, this case qualifies as a definite paraneoplastic neurological syndrome. The patient had a non classical neurological syndrome with well characterised onconeural Ab and a cancer that was found within few months of the neurological disorder.

The patient had some characteristic clinical manifestations described with anti-Hu syndrome, as limbic encephalitis<sup>5</sup>. She also had rarer features of the syndrome. Bilateral tonic pupils have been described in association with anti-Hu Ab, but seem relatively rare. Unilateral presentation is more common. Autonomic failure at large, including abnormal pupillary responses, has been described in 28% of patients, but the prevalence varies between different series<sup>5</sup>. Our patient had a man-in-the-barrel syndrome attributable to her paraneoplastic syndrome. Lower motor neuron involvement, with progressive

proximal upper arm weakness spreading distally, has been reported in 14 % of patients with anti-Hu Ab<sup>5</sup>.

There is one case in the literature of a malignant mixed müllerian tumor (carcinosarcoma) associated with anti-Hu paraneoplastic sensory neuronopathy<sup>2</sup> resistant to pharmacological and surgical treatments, with another case of cervical cancer with anti-Hu Ab, cerebellar ataxia and sensory neuropathy having been reported<sup>3</sup>.

This case exemplifies once again that the spectrum of cancers associated with anti-Hu can be enlarged to include gynaecological tumors.

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## TO THE EDITOR

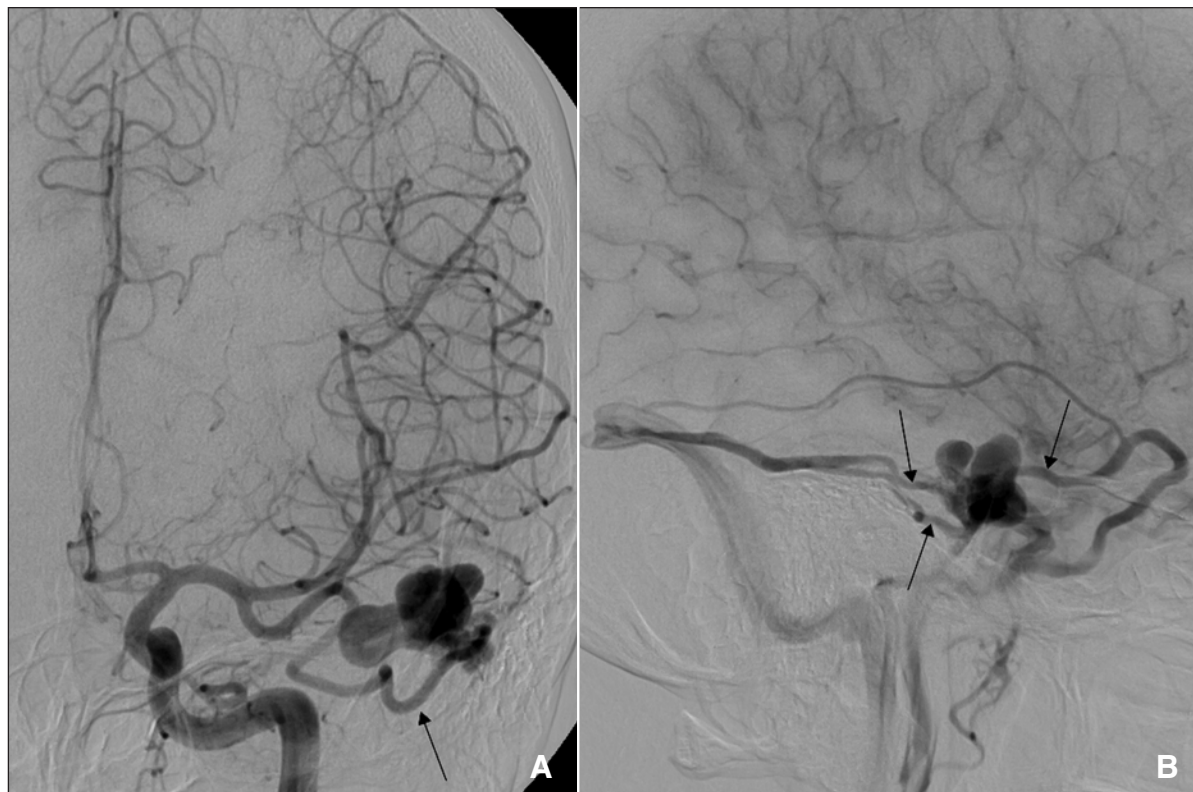
### Spontaneous Occlusion of the Temporal AVM Associated With Tinnitus

Arteriovenous malformations (AVMs) of the brain were traditionally considered congenital abnormalities. However, there have been a fair number of case reports describing a de novo origin and an eventual spontaneous regression of the AVMs suggesting a dynamic nature to some of these lesions. These lesions can be occult at the early stages, grow over time and later regress. According to Lasjaunias<sup>1</sup>, the origin of AVMs might be explained by continuous vascular remodeling which is triggered by a provocative event altering the normal vascular development. Pulsatile tinnitus is a recognized clinical presentation of cerebral AVMs. The spontaneous occurrence of tinnitus in patients, with an otherwise clinically silent AVM, supports the fluctuating growth of these malformations. These malformations can further lead to more critical changes in the cerebral hemodynamics. To our best knowledge, the spontaneous

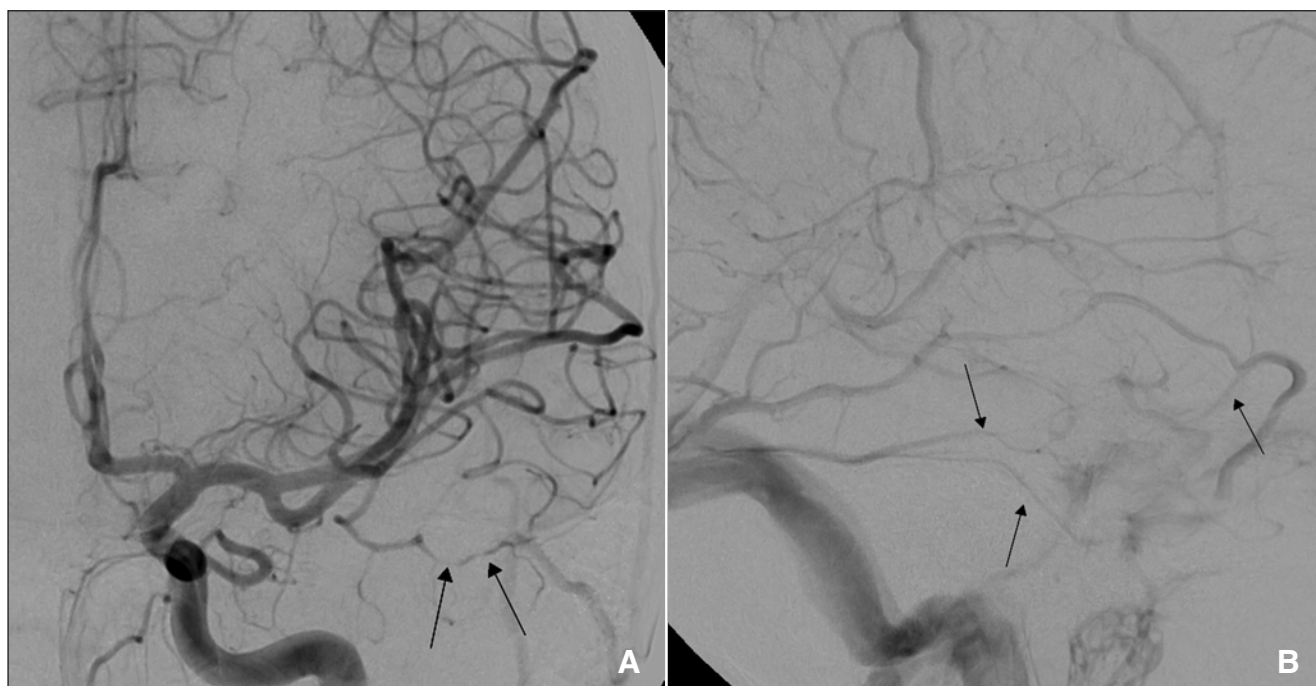
regression of an AVM causing a transient tinnitus has not been reported previously.

## CASE REPORT

A 21-year-old female presented with a six month history of left sided pulsatile tinnitus and was diagnosed with a 3 cm superficial left temporal AVM on magnetic resonance imaging (MRI). She was consequently evaluated with a conventional angiogram which demonstrated a solitary feeding artery arising from one of the M2 branches of the left MCA (Figure 1A) and three cortical veins draining the malformation into the left inferior petrosal and sigmoid sinuses (Figure 1B). An elective embolization was chosen as the method of treatment. Two months later, on the day of the procedure, the repeat angiogram failed to demonstrate patency of the nidus of the malformation. Several segmental areas of stenosis of the feeding artery and occlusion of the proximal segment of the largest draining vein were observed (Figure 2A and 2B). The patient's symptoms resolved, and a follow up MR performed one month later confirmed the thrombosis of the malformation.



**Figure 1:** A) Frontal projection of the selective right internal carotid angiogram shows a small left temporal AVM with a single feeding artery (arrow) arising from the M2 segment of the left MCA. B) Lateral angiographic projection outlines three cortical veins (arrows) providing the outflow into the sigmoid and inferior petrosal sinuses.



**Figure 2:** A) Two-month follow-up angiographic image with a magnified region of interest demonstrates resolution of the malformation and segmental areas of stenosis affecting the supplying artery (black arrow). B) Lateral projection shows absence of flow within the malformation and proximal segments of the draining veins. More distal segments remain patent due to collateral circulation (arrows).

**DISCUSSION**

The incidence of spontaneous obliteration of cerebral AVMs is fairly rare and is estimated to occur in only 0.8% -1.3% of cases<sup>2</sup>.

Multiple theories have been proposed to explain this spontaneous regression. The potential mechanisms for this occurrence can be divided into three groups based on whether they affect the arterial inflow, venous outflow or both. A previously reported explanation proposes that the intracranial hemorrhage with its subsequent mass effect and decreased blood to the lesion can lead to thrombosis of the AVM. Other causes of increased intracranial pressure, such as an intracranial mass and hydrocephalus, have also been associated with this phenomenon. The regression of AVMs in the absence of increased intracranial pressure has been much less frequent. Panciani et al provided an extensive literature review of 93 patients out of which only 27 cases of spontaneous occlusion of AVMs were not associated with hemorrhage or tumor<sup>3</sup>.

Another possible theory suggests that the obstruction of the arterial inflow can also cause spontaneous regression of the malformation. This obstruction may be secondary to a local thrombotic process and/or an embolus originating from the heart or proximal artery. High flow angiopathy, which has been evaluated in pathologic studies and experimental animal models, has been postulated as another potential mechanism which can cause stenosis and thrombosis of the vessels involved<sup>4</sup>. Lastly, the obstruction of the outflow vessels may also lead to a complete thrombosis of the entire lesion as described by Sawlani et al<sup>5</sup>. Small AVMs, like the 3cm AVM with a solitary feeding artery and three draining veins in our patient, which have few feeding arteries and draining veins are more likely to regress spontaneously than larger lesions recruiting multiple vessels<sup>2</sup>.

In our particular case, the absence of any known risk factors for thromboembolism, along with the angiographic demonstration of multisegmental narrowing of the feeding artery suggest the high flow angiopathy theory as the more likely plausible cause.

In conclusion, spontaneous occlusion of the AVM is a rare event which seems to be the result of a complex interplay between host factors and the malformation architecture. The development of tinnitus in a 21-year-old female secondary to an underlying AVM, which later regresses, helps confirm the dynamic nature of these lesions. We believe that, in this particular case, the progressive changes in the hemodynamics of the malformation due to high flow angiopathy help explain the initial development of the pulsatile tinnitus and its subsequent spontaneous resolution.

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