Autoimmune Endotheliopathy in the Inner Ear: Audiologic Abnormalities of SS

The third component of the SS clinical triad is inner ear involvement---involvement of the cochlea and the vestibular apparatus (semi-circular canals). Again, it is an autoimmune endotheliopathy in the microvascular of the inner ear that is the problem. This endotheliopathy leads to narrowing of the channel within the tiny blood vessels that feed the cochlea and vestibular apparatus, and the resultant decreased blood flow and decreased delivery of oxygen and nutrients causes ischemic injury to these tissues.

Figure 32 shows the anatomy of the inner ear. Because it is the blood supply to the tip of the cochlea that is apparently most vulnerable, and because the tip of the cochlea is responsible for hearing sounds of low frequency, the autoimmune endotheliopathy of SS primarily causes low frequency sensorineural hearing loss. Loss at other frequencies, however, can also occur. The hearing loss of SS can be on one side, both sides, equal, or unequal. It can develop suddenly, or more gradually and progressively. It can be so profound as to require cochlear implants.

Although the hearing loss can certainly affect the patient’s speech, the patient’s speech can also be affected by a different problem---ischemic injury to the speech center in the brain (i.e. dysarthria, “slurred speech”, due to autoimmune endotheliopathy in that location). So, in SS a patient’s speech difficulties can stem from injury to either the cochlea, or the brain, or both, and in varying combinations. Likewise, poor speech discrimination can be due to either the hearing loss alone, or to ischemic injury to the speech discrimination center in the brain, or to both, in varying combination.

The hearing loss is often accompanied by tinnitus (“ringing” or “humming” in the ears). This tinnitus is often “roaring” and can be extremely distressful and upsetting to the patient. Typically, the tinnitus, even when initially “roaring,” gradually diminishes in intensity, but this can take several months before it becomes truly tolerable, and then it often persists for a long time, usually at a much more tolerable level.

When the vestibular apparatus is involved, the patient is likely to note dizziness (a “spinning sensation,” “whirling” vertigo). This dizziness/vertigo is different from a second potential cause of loss of balance that can also occur in SS, due to autoimmune endotheliopathy in the microvasculature of the cerebellum. So, again, there are at least two reasons for “poor balance” in SS---ischemic injury to the vestibular apparatus and ischemic injury to the cerebellar tissue---and both can occur in the same patient. Furthermore, weakness from hemiparetic injury can also cause poor balance.