Acquired Auditory-Tactile Synesthesia

Synesthesia is the automatic elicitation of conscious perceptual experiences by stimuli not normally associated with such experiences (eg, tasting words, hearing colors). The stimuli that induce synesthesia can be either cognitive (eg, thinking of a number) or sensory (eg, listening to music). Synesthesia can be either developmental in origin (present throughout the life span, with a hereditary component) or acquired. Whereas developmental cases tend to be either of the cognitive-sensory or sensory-sensory types, acquired cases tend to be limited to the sensory-sensory types. (Note: I use the convention of placing the stimulus that elicits the experience before the hyphen.) This could reflect the fact that the onset of synesthesia predates the learning of cultural knowledge (eg, words, letters, numbers) in developmental but not in acquired cases. Acquired cases of synesthesia have not been extensively documented, and Ro et al. study makes an important contribution to this emerging literature.

Most previous cases of acquired synesthesia have arisen as a result of sensory deafferentation in the visual modality resulting in acquired visual synesthesias such as sound-vision and touch-vision synesthesias. These cases typically have peripheral damage to the visual pathways (eg, retinal degeneration or optic nerve damage), although one case of sound-vision synesthesia after midbrain tumor is reported. Ro and colleagues’ case is unique in having a discrete neurological lesion, in this instance, to the right ventrolateral thalamus. It is also unique in that tactile sensations are elicited from sounds.

The facts of the case are as follows. At 12 months after onset, a neuropsychological investigation demonstrated the rare condition of tactile and visual antiextinction (ie, improved ability to detect a contralesional stimulus when accompanied by an ipsilesional stimulus of the same modality). Ro and colleagues speculate that this may be caused by compensatory plasticity in corticocallosal pathways. The patient also had decreased somatosensory functions on the left (contralesional) side of her body. A magnetic resonance imaging scan and diffusion tensor imaging analysis at about 15 months showed disorganized white matter in the lesioned hemisphere. It is unclear whether the disorganized subcortical connections themselves cause the synesthesia, or whether it is due to other compensatory corticocortical plasticity (see later). The synesthesia itself lasted for many years and was reassessed at 6 years after onset. The tactile percepts were typically experienced on the left upper part of the body (including hands and arm), were simple in nature (eg, tingles or pressure rather than shapes), and were generally consistent over time.

Given that the synesthesia and the earlier impairment in sensory functioning occurred within the same domain (ie, tactile sensations on the left), it resembles previous cases of acquired visual synesthesia after visual impairment (even though the lesion was different in nature). As such, I am inclined to attribute the synesthesia to compensatory cross-modal plasticity rather than to loss of thalamic input in particular (although further evidence concerning the role of the thalamus in synesthesia is clearly needed). The slow time course of onset of the synesthesia also resembles other cases and is more consistent with plasticity rather than unmasking of pre-existing pathways. Unmasking could be a viable mechanism for some cases who report synesthesia after only a few days, for healthy blindfolded participants who report visual experiences after a few days, and for drug-induced synesthesia.

The specific nature of the synesthesia is also intriguing. The modality of the experiences (ie, touch) is perhaps predictable from the sensory deficit. But why do sounds act as the inducing modality? One simple answer is that there is nothing special about sounds, and other sensory combinations will be documented when new cases of acquired tactile synesthesia come to light. However, other explanations can be considered. Developmental types of synesthesia may be constrained by the neuroanatomic proximity of functional regions, such that color perception and grapheme recognition are neuroanatomically close (creating grapheme-color synesthesia), whereas spoken language, rather than graphemes, is linked to synesthetic experiences of taste. This “adjacency principle” is assumed to reflect a genetic bias that affects brain development in naturally occurring cases, so it is not obvious whether such a constraint will apply to acquired cases. The fact that the tactile sensations were felt on the upper (rather than lower) body is, however, consistent with this. The region of somatosensory cortex representing the face, hands, and arms lies closer to the auditory cortex than that representing torso, legs, and feet. However, one might expect, under an adjacency account, that gustatory-tactile synesthesia would predominate over auditory-tactile synesthesia. (Cytowic and Wood report such a developmental case). An alternative is that there already exist direct auditory-tactile pathways in
Primary Central Nervous System Vasculitis: Progress and Questions

Vasculitis limited to the central nervous system (CNS) represents a rare and serious disorder that over the past several decades has increasingly been considered in the differential diagnosis of patients with unexplained neurological manifestations. This condition, known by numerous labels including primary angiitis of the central nervous system (PACNS),1,2 isolated angiitis of the CNS,3 and primary vasculitis of the CNS,4 represents a major diagnostic and therapeutic challenge. This challenge is magnified by the numerous unanswered questions regarding the pathogenesis, optimal diagnosis, treatment, and prognosis of PACNS. In this issue of the Annals, Salvarani and colleagues5 provide a retrospective analysis of the largest cohort of such patients ever reported. The accrual of this cohort represents no small feat, requiring the formidable resources of the Mayo Clinic over two decades, reaffirming that even in centers such as this, which excel at attracting and evaluating rare, complex disorders, the frequency of encountering PACNS amounts to only a handful of such patients per year. Given that up until 1980, there were only a few dozen of such reported cases in the world literature, this single center investigation of 101 patients adds formidably to the existing body of clinical experience, whereas at the same time raising a series of questions regarding future efforts to further understand this disease.

How has this series furthered clinicians’ understanding of PACNS? This report reaffirms that vasculitis confined to the CNS, regardless of whether diagnosed by biopsy or angiography, is most certainly a heterogeneous collection of disorders. Similar to previous reports,6 differences, although subtle, were detected between angiographically and histologically confirmed cases with trends for more female individuals and between angiographically and histologically confirmed cases with trends for more female individuals and benign spinal fluid results in the angiographically defined group. For diagnosis, this study found that brain magnetic resonance imaging was abnormal in 97% of patients, reaffirming that brain magnetic resonance imaging has a high negative predictive value in the diagnostic assessment, thereby providing reassurance for clinicians assessing such patients. Although, as candidly noted by the authors, the retrospective study design precludes assessment of the relative efficacy of the various therapeutic approaches, this report suggests that a considerable number of patients may respond well with glucocorticoid therapy alone. In addition, this

the human brain that get strengthened as a result of neurological or sensory impairment. Further evidence, including from other cases, is needed to adjudicate among these three different alternatives.

It is also unclear why some patients develop symptoms of synesthesia, whereas others do not. Is it due to the type of lesion or to pre-existing differences in the patients themselves? Currently, acquired synesthesia appears to be rare, but I suspect that this may change now that clinicians (and patients) are more familiar with this concept. Further studies in this area will undoubtedly lead to important insights into cross-modal plasticity and the organization of multisensory processes more generally.

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References

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