Intraneural ganglion

To the Editor: The recent report of a so-called hypoglossal intraneural cyst by Nonaka et al. is a remarkable one (Nonaka Y, Grossi PM, Filomena CA, et al: Unilateral hypoglossal nerve palsy caused by an intraneural ganglion cyst. Case report. J Neurosurg 113:380–383, August, 2010). Nonaka et al. did not identify a joint connection. They found the tumor “to originate from the hypoglossal canal” (a site without synovium), concluding that the “tumor originated from the hypoglossal nerve rather than the atlantooccipital joint.” They proposed a different mechanism for the formation of their case, one contrary to the unifying articular (synovial) theory. Their paper suggests to us 2 viable explanations: 1) this case would be the first bona fide exception to the articular (synovial) theory, or 2) it follows the rule. We are grateful to these authors for sharing the complete set of MR images with us after their publication in an effort to not only promote but also advance science.

Recently we extrapolated the principles of the articular theory from the appendicular skeleton to the axial spine, demonstrating its versatility in explaining extraneural and intraneural cysts. We had previously excluded hypoglossal cysts from our analysis of juxtafacet cysts due to the difficulty of confirming the specifics about previous cases—namely, the intraneural or extraneural nature of these cysts was not always substantiated, remained in doubt, or was controversial. In fact, the previous examples of hypoglossal cysts included in the paper by Nonaka et al. have all been shown or presumed to have an atlantooccipital joint connection. That juxtafacet cysts develop on occasion at the craniovertebral junction appears to be no surprise because both the atlantooccipital joint and atlantoaxial joint are synovial joints; nor is their potential for compression of neighboring nerves (including the hypoglossal nerve) or the spinal cord unanticipated.

The articular connection now established in the case published by Nonaka et al. solidifies the explanation for the formation of these cysts. Unfortunately, the imaging parameters are not detailed enough to allow us to delineate the propagation. Creativity and finesse will be needed to explain an intraneural occurrence: as the hypoglossal nerve, a pure motor nerve, does not have an articular branch to the upper cervical spine, an immediate explanation is not available. We previously postulated that the ansa cervicalis could provide the answer to the link between the hypoglossal and the C-1 and C-2 nerves, a neural communication that could explain the anatomy and perhaps the pathology. Further investigations with high-resolution thin-slice images will undoubtedly unveil the propagation patterns in future patients with this rare condition.

The case presented by Nonaka et al. illustrates the difficulty, even for master surgeons, to establish the diagnosis of an atypical juxtafacet cyst and to identify its joint connection, either preoperatively on imaging or at operation, when it occurs in an unusual location. Just as the cyst itself was originally not identified 7 years earlier when it measured 3–4 mm, the pedicle of these cysts is often small (measuring only 1 mm in this case) and can
easily be missed. Likewise, this case illustrates the ease of claiming that the articular theory does not apply without providing definitive evidence to disprove it.

Understanding the pathogenesis of these cysts provides insight into how best to treat the pathology. We are learning that in intraneural and extraneural ganglia the joint is the primary problem; the cyst, which is the secondary problem, and often the more imminent one, need not always be directly addressed if the primary problem is adequately treated. We also know that incorrectly treated, these cysts can recur at alarming rates. Only through such collaborations will we be able to further elucidate the mechanisms and solve these types of clinically relevant problems.

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References

Response: We thank Dr. Spinner and his coworkers for their comments. We are very much aware of their elegant work defining the origin of ganglion cysts in the peripheral nerves. In most cases, the affected nerve has a branch to an adjacent joint. Their papers were very much on our minds when we discovered the cyst at the time of surgery. Because we did not expose the entire nerve from its intracranial origin through its extracranial course, we can not say with certainty that there was not an imperceptible stalk that traveled from the joint along the nerve and then blossomed in the proximal hypoglossal canal. On dissecting the cyst from the fascicles of the hypoglossal canal and the hypoglossal nerve no such stalk was noted (Figs. 1 and 2).

The application of Dr. Spinner’s theory to this case is significant. If Dr. Spinner’s theory is correct, the cyst should recur as the connection with the joint’s synovium is still intact. We know, as does Dr. Spinner, of a description of an articular branch from the hypoglossal nerve to the atlantooccipital joint. The possibility of an ectopic synovial cyst can not be ruled out in this case.

Fig. 1. The inferior portion of the tumor was adherent with the fascicle of the hypoglossal nerve (asterisk). PICA = posterior inferior cerebellar artery. XI = cranial nerve XI.

Fig. 2. Illustration demonstrating the relationship between the tumor and surrounding structures. The tumor was located at the level of the hypoglossal canal with compression of the hypoglossal nerve downward. A-O joint = atlantooccipital joint. Modified with permission from Fukushima T: Manual of Skull Base Dissection, ed. 2. AF-Neurovideo, 2004.