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EDITORIAL

An historical perspective on ulnar intraneural ganglion cysts and their joint origins

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The case report by Xu et al. [24] of an ulnar intraneural ganglion at the elbow is an important one. The topic should be of interest to hand surgeons as it pertains to the classic anatomy and the rich heritage of our discipline. These authors describe an old problem but employ a modern solution. They recognized that the primary pathology was the elbow joint and not the intraneural cyst. At surgery they identified and disconnected the articular branch connection and decompressed the cyst. Their patient had an excellent clinical result and no recurrence. Their clinical case offers us the opportunity to give an historic perspective on the progress made over the past two centuries with intraneural ganglia: the pathogenesis of this entity, once controversial, is becoming clear; operative interventions which were intricate are being simplified; surgical outcomes, previously suboptimal, are improving; and recurrences, once as high as 30 %, are becoming much less common—potentially a thing of the past.

In 1810, Beauchêne fils (a French anatomist and surgeon and student of Dupuytren) described the unusual finding of a *kyste cubitale dans un homme environs quarante ans* (an ulnar nerve cyst in a man about 40 years of age) [3]. This short description is that of the first known intraneural ganglion cyst, which happened to be involving the ulnar nerve at the elbow.

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Intraneural cysts were considered a "curiosity" from Bertrand in 1837 [2] through Seddon in 1975 [14] to Birch in 1998 [5]. Several theories—such as degenerative (de novo), articular, tumoral, and extraneural intrusion—emerged to explain the formation of these cysts: the first two became the popular ones, though neither explained all cases and observations satisfactorily.

In 2003, the unifying articular (synovial) theory for intraneural ganglion cysts was proposed based on an extensive literature review and a large retrospective multicenter study. Developed on the most common site (the peroneal nerve at the fibular neck arising from the superior tibiofibular joint) [15, 16], this unifying theory could be extrapolated to intraneural cysts at other sites [18], including those affecting the upper limb [22]. It offered a single, anatomic explanation that could explain all reported cases at all locations, their phenotypic similarities and apparent outliers. According to this unifying theory, intraneural ganglions form from neighboring (often degenerative), synovial joints; joint fluid dissects within an articular branch and extends to the parent nerve. Propagation occurs along a path of least resistance and is influenced by pressure and pressure fluxes. The theory stated that the joint connection unidentified in the 60 % of reported cases had not been recognized. The joint connection has been substantiated in more than 100 consecutive cases prospectively reviewed and treated by our group. Many of these cases were cases of others (including a previously published case of an ulnar intraneural ganglion cyst) [6, 22] where joint connections were not previously identified, and the ganglions had recurred. The use of high-resolution imaging (including magnetic resonance arthrography) and post-processing three-dimensional reconstruction has helped us identify pathognomonic features to diagnose the cysts and delineate the joint connection to the point where this type of technology, while revealing, is not really needed for diagnosis. Expecting the joint connection to be there is the first step in finding it—either

preoperatively or intraoperatively. Other groups have corroborated the joint connections at many sites and have endorsed the theory [1, 8, 9, 12, 13, 23], including Birch in 2011 in the most recent edition of his classic book [4]. The unifying articular theory can be translated directly to clinical outcomes: successful treatment entails disconnection of the cyst from the

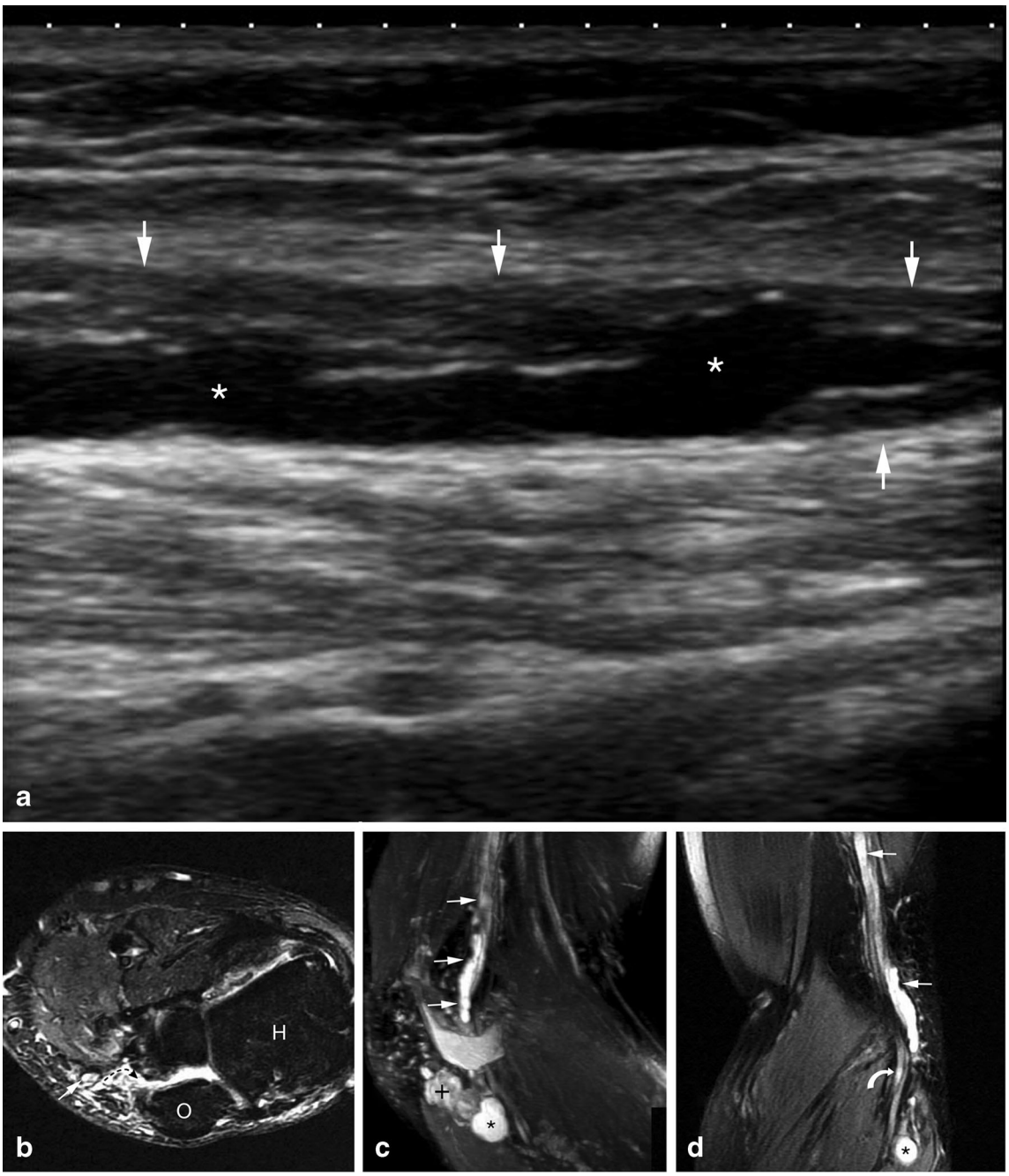

Fig. 1 A 59-year-old man presented with a 2-year history of severe ulnar neuropathy and was found to have an ulnar intraneural ganglion arising from the elbow joint on ultrasound (**a**) and magnetic resonance imaging (**b**–**d**). **a** Longitudinal ultrasound of the right upper arm shows hypoechoic intraneural cyst (*asterisks*) extending along the ulnar nerve (*arrows*). The tubular mass extended 12 cm from the elbow to the mid-arm. The nerve is slightly hypoechoic overall, consistent with edema in the nerve. **b** Axial short-tau inverted recovery image just proximal to the elbow joint shows an intraneural cyst within the ulnar nerve within the cubital tunnel (*arrow*) with the connection to the elbow joint (*dashed arrow*) well seen. *O* olecranon, *H* distal humerus. **c** Sagittal oblique maximum intensity projection from a short-tau inverted recovery data set showing the proximal extent of the intraneural cyst within the ulnar nerve (*arrows*), an extraneural soft tissue cyst just distal to the elbow (*asterisk*), and an intraosseous cyst in the proximal ulna (*cross*). The individual joint connections are not seen on this image. **d** Sagittal oblique maximum intensity projection image from a short-tau inverted recovery data set shows intraneural cyst within the ulnar nerve extending both proximally (*arrows*) and distally (*curved arrow*) from the origin at the elbow joint (not shown). Note also extraneural cyst (*asterisk*)

joint; when this is done, intraneural recurrence can be eliminated. Note that intraneural recurrence is frequently seen in peroneal nerve cases where the articular branch is not exposed but rarely in upper limb cases where the articular branch is often inadvertently resected during dissection. If the articular branch connection is not addressed, recurrence could and often did occur with careful, long-term follow-up, although it may not be recognized or reported. The cyst can also be decompressed to expedite resorption. More aggressive treatments, such as resection of the cyst or the nerve itself, the latter still being performed [7], are not necessary.

In 2008, the primary author (RJS) sought to identify and verify the correct citation of the original Beauchêne specimen [19]. What is more, the specimen, which had been maintained in an anatomic museum for several decades, survived the wars and was now displayed on the shelves in the Musée Dupuytren in Paris. A trip to France allowed reexamination of the specimen. The finding of a previously unrecognized cystic articular branch connection to the elbow in this specimen provided strong historic evidence to support a joint origin for this first case of an intraneural cyst and in all intraneural cysts that were to follow.

The case of Xu et al. [24] is a modern-day recapitulation of Beauchêne's case. The similarities are not coincidental and do not end there. The magnetic resonance imaging findings in the case of Xu et al. were classic for a joint-connected intraneural cyst (though misdiagnosed by the authors prospectively) and reminiscent of those in the first known case of an ulnar intraneural cyst in which magnetic resonance imaging revealed the joint connection to the elbow [10]. For this commentary, we reinterpreted that intraneural case and identified coexisting extraneural soft tissue and intraosseous cysts derived from a degenerative joint (Fig. 1). The constellation of findings is easily understandable because of the shared pathogenesis of these interrelated cysts.

Review of the literature has uncovered approximately 500 cases of intraneural ganglions, including more than 55 cases of ulnar intraneural ganglions at the elbow (38 of which were summarized in a recent publication) [22]. The ulnar nerve at the elbow is the second most common site for intraneural ganglions and is the prototype for intraneural cysts in the upper limb. Overall, one third of the ulnar intraneural ganglia at the elbow had recognized joint connections. Based on the layers of research, we strongly believe that the others were unrecognized.

But history and historical evidence for these ulnar nerve cysts do not end here—at the elbow. In 1884, the first described joint-related intraneural cyst, an ulnar intraneural ganglion at the wrist, was found in a cadaveric specimen that could be traced to Sir Anthony Bowlby (the noted British anatomist, surgeon and author of an early book on peripheral nerve injuries) [20]. A recent review of these clinical cases concluded that these cysts were joint-related [21]. In 1961, Parkes (an orthopedist and peripheral nerve surgeon practicing in Scotland), in a paper on peroneal intraneural ganglion cysts, informally provided information in which he identified the

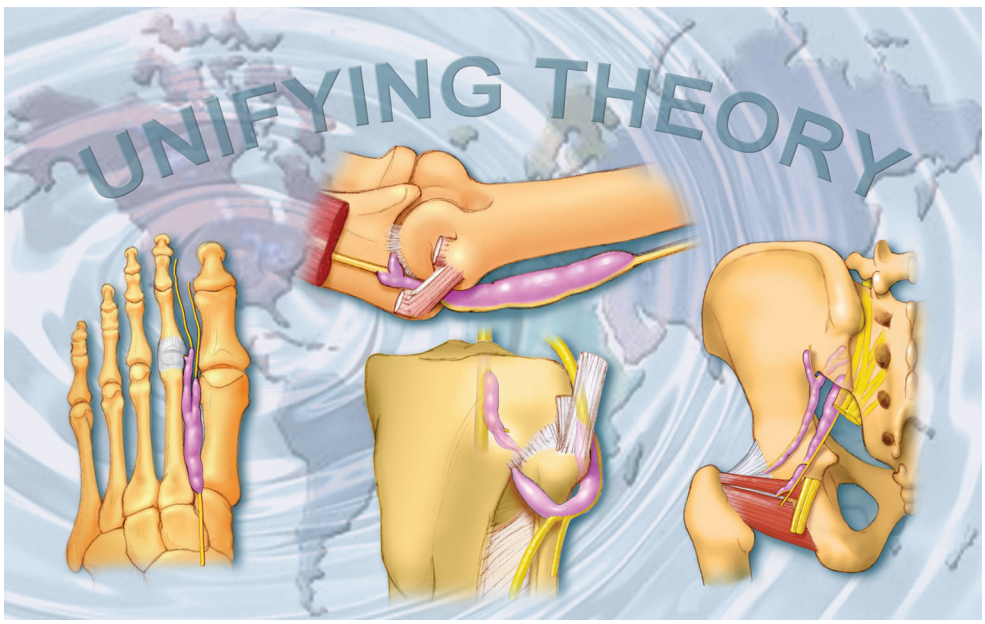

Fig. 2 The unified articular (synovial) theory is gaining global acceptance. It was put forth to support the most common example, the peroneal nerve at the fibular neck. It has been substantiated for other intraneural ganglion cysts—from the hip to the toe. The drawing of the ulnar nerve example at the elbow was based on the reinterpretation of Beauchêne's specimen [19]. This original case is remarkably similar to the clinical case reported by Xu et al. [24]. The drawing shows the propagation pathway. The pulsation from increased intra-articular pressures inferred in the drawing can be appreciated on the ultrasound of the featured clinical case of an ulnar intraneural ganglion at the elbow (see Fig. 1a) (with permission, Mayo Foundation 2013)

first case of an adventitial cyst within the ulnar artery which was (wrist) joint-connected. He suggested a similarity of adventitial cysts to intraneural cysts [11]. A recent review proved him right [17]. The articular theory could be extended to adventitial cysts (i.e., arteries or veins), potentially ending similar controversies on that related entity. Adventitial cysts act the same way whether at the common site (i.e., the popliteal artery at the knee via the middle genicular artery) or at rare sites [17]. The single anatomic explanation for this entity is equally appealing and intuitive: capsular vessels run next to nerves (i.e., a neurovascular bundle).

This historical perspective provides insights into the controversies regarding the pathogenesis and treatment of ulnar intraneural cysts at the elbow and those throughout the body. The articular branch connection of these cysts is fundamental to these cases and is consistently there, recognized or not. The joint connection may be subtle and is easy to miss with routine imaging and at operation, especially for those not familiar with this rare entity. This report by our international colleagues highlights the increasing global awareness of the articular (synovial) theory (Fig. 2). We acknowledge that global acceptance may be years away, we hope, catalyzed by technological advances resulting in more rapid dissemination of information. We need to learn from the past, which has enlightened us in the present and, no doubt, will illuminate the way for further advances in the future.

Conflict of Interest Robert J. Spinner declares that he has no conflict of interest.

Srinivasan Harish declares that he has no conflict of interest.

Kimberly K. Amrami declares that she has no conflict of interest.

Statement of Human and Animal Rights There is no violation of human and animal rights.

Statement of Informed Consent There is no informed consent as no human subjects are involved.

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