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CASE REPORT

CASE REPORT: DIAGNOSIS AND MANAGEMENT OF CARPAL TUNNEL SYNDROME IN A PATIENT WITH OLIGODACTYLY

OPIS PRZYPADKU: DIAGNOSTYKA I POSTĘPOWANIE W ZESPOLE CIENI NADGARSTKA U PACJENTA Z OLIGODAKTYLIĄ

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ABSTRACT
Carpal tunnel syndrome (CTS) is a frequent neuropathic condition that can result in numbness, pain, and tingling sensations in the fingers and hand. It often affects individuals who have specific risk factors, such as injuries, pregnancy or certain medical disorders.

In this case report, we describe a patient presenting with symptoms of carpal tunnel syndrome (CTS), along with radial longitudinal deficiency, which represents a spectrum of upper limb dysplasias and hypoplasias. The patient exhibited unilateral thumb and radial artery hypoplasia, with the ulnar artery being the exclusive source of blood supply to the hand. Initially, the patient reported mild CTS symptoms that gradually worsened, impairing her daily activities. An ultrasound confirmed the diagnosis, and surgical release of the carpal tunnel was performed. Following surgery, the patient reported significant improvement in her symptoms, with her hand and finger strength and sensation returning to baseline.

The aim of this case report is to enhance the knowledge and skills of orthopedic practitioners by presenting a detailed account of the diagnosis and management of carpal tunnel syndrome in a patient with oligodactyly and an unique anatomical variation of hand vascularity. We discuss the topic of radial longitudinal deficiency and typical anatomical variations in human hand vascularity.

Keywords: Carpal tunnel syndrome, oligodactyly

STRESZCZENIE
Zespół cieśni nadgarstka (ZCN) jest częstym schorzeniem neuropatycznym, które może powodować drętwienie, ból i uczucie mrowienia w palcach i dłoni. Często dotyka osób z określonymi czynnikami ryzyka, takimi jak urazy, ciąża lub niektóre schorzenia.

W niniejszym opisie przypadku przedstawiamy pacjentkę z objawami zespołu cieśni nadgarstka (ZCN) oraz niedoborem promieniowym podłużnym, który reprezentuje spektrum dysplazji i hipoplazji kończyny górnej. U pacjentki stwierdzono jednostronną hipoplazję kciuka oraz tętnicy promieniowej, przy czym tętnica łokciowa była jedynym źródłem zaopatrzenia dłoni w krew. Początkowo pacjentka zgłaszała łagodne objawy ZCN, które stopniowo nasilały się, utrudniając jej codzienne czynności. Badanie ultrasonograficzne potwierdziło diagnozę, a pacjentka przeszła chirurgiczne uwolnienie cieśni nadgarstka. Po operacji zgłaszała znaczną poprawę objawów, z powrotem siły i czucia w dłoni i palcach do stanu wyjściowego.

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Celem tego opisu przypadku jest poszerzenie wiedzy i umiejętności ortopedów, poprzez szczegółowy opis diagnozy i leczenia zespołu cieśni nadgarstka u pacjentki z oligodaktylią i unikalnym wariantem anatomicznym unaczynienia ręki. Omawiamy również zagadnienie niedoboru promieniowego podłużnego oraz typowe warianty anatomiczne unaczynienia ręki.

Słowa kluczowe: zespół cieśni nadgarstka, oligodaktylia

Introduction

Carpal tunnel syndrome (CTS) encompasses a spectrum of clinical manifestations that result from median nerve entrapment within the carpal tunnel. Patients typically report experiencing pain and paresthesia, and less frequently, weakness, in the distribution of the median nerve. CTS is the most commonly observed compressive focal mononeuropathy in clinical practice, with an estimated prevalence of 1 to 5 percent in the general population. Notably, it is more prevalent in females (Pourmemari *et al.,* 2018 and Atroshi *et al.,* 1999). Multiple factors increase the risk of developing CTS, including diabetes mellitus (Albers Leach *et al.,* 1996 and Leach *et al.,* 1968), arthritis (Shiri 2016), thyroid disorders (van Dijk *et al.*, 2003), pregnancy (Padua *et al.,* 2001), and wrist trauma (Pope and Tang 2018) (Schnetzler 2008). The contribution of repetitive hand and wrist use and workplace factors to the onset of CTS is an area of ongoing debate among experts (Padua *et al.,* 2016; Shiri and Falah-Hassani, 2015; Mediouni *et al.,* 2014).

Radial longitudinal deficiency is a condition characterized by a spectrum of dysplasias and hypoplasias in the upper limb, primarily affecting the radial aspect of the forearm, wrist, and hand. It involves bony abnormalities in the thumb and radius, as well as deficiencies in muscles, nerves, vessels, and joints, leading to significant functional impairments in the upper extremity (Bhat and Acharya, 2020; Maschke *et al.,* 2007; Ekblom *et al.*, 2013). This condition may be associated with other medical conditions such as TAR (thrombocytopenia absent radius) syndrome, Fanconi's anemia, Holt-Oram syndrome, and the VATER (vertebral anomalies, anal atresia, tracheoesophageal fistula, esophageal atresia, renal agenesis) or VACTERL (vertebral anomalies, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal agenesis, and limb defects) association, which involve various concomitant anomalies in different organ systems (Maschke *et al.,* 2007; Ekblom *et al.*, 2013; Forman *et al.*, 2020).

In this article, we present a case of CTS in a patient with radial longitudinal deficiency. Although our literature search did not yield strong evidence that supports a correlation between congenital hand anomalies and the development of CTS, it is plausible that such a relationship exists.

Case presentation

A 36-year-old female with a history of left thumb hypoplasia presented to the clinic with complaints of pain, numbness, and tingling in her left hand and fingers. Her symptoms had started in April/May of 2022, and had gradually worsened over time, affecting her daily activities such as holding a cup, combing her hair, and using her phone. She also reported frequent episodes of hand numbness and tingling, as well as decreased hand grip strength, resulting in objects slipping out of her hand. Her pain was most severe in the summer of 2022, and was worse at night and on raising her arm. Her symptoms improved when she lowered her hand. She had been taking various pain medications, including ibuprofen, acetaminophen, dexamethasone, and ketoprofen, with ketoprofen being the only medication providing relief.

The patient had a history of oligodactyly and she had no other medical conditions and did not take any regular medications.

The patient did not present with any additional disorders often associated with radial longitudinal deficiency, including anemia and various organ anomalies. She denied any history of trauma or repetitive hand motions. During pregnancy, her mother was living in Poland when the Chernobyl disaster occurred, which might have contributed to the development of congenital abnormalities of the patient. The patient had sought medical attention in July/August of 2022, and underwent an electromyography (EMG), which was suggestive of mild nerve compression but not CTS. The diagnosis of CTS was made in November 2022 after an ultrasound of the hand and wrist showed evidence of median nerve compression at the wrist (Figure 1, 2). Furthermore, the ultrasound revealed the absence of the left radial artery in the patient, with the ulnar artery being the sole source of blood supply to her hand. Nevertheless, the patient was qualified for a decompression surgery of the compressed nerve.

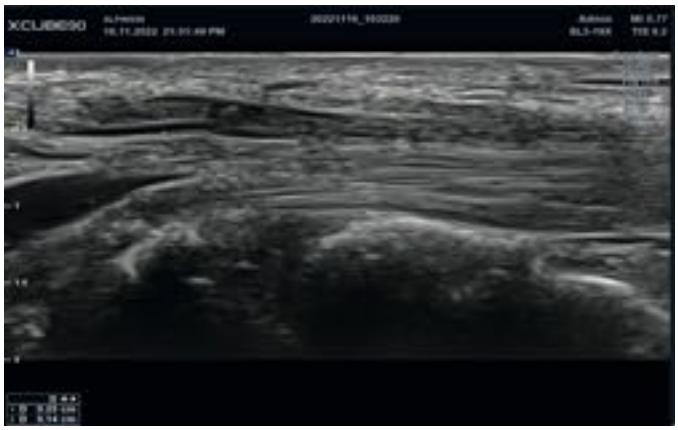

Figure 1. A longitudinal cross-section of the median nerve. The nerve is narrowed and appears more echogenic below the transverse ligament. There is an edema in the proximal portion of the nerve, which is characterized by thickening, reduced echogenicity, and disrupted fibrillar echostructure

Management and outcome

The procedure performed to this patient was a release of the left median nerve, done under Esmarch's tourniquet for ischemia control (6 minutes ischemia time for nerve release). The arcuate incision was performed above the transverse ligament. The soft tissues were dissected until reaching the transverse carpal ligament. A grooved probe was then inserted under the ligament, covering the nerve and lifting the superficial tissues, and the ligament was cut posteriorly, partially subcutaneously, along with a part of the forearm fascia, approximately 1.5 cm proximally from the wrist flexure line. The thickened ligament was found to be constricting the nerve, which was also thickened proximally. Hemostasis was checked, and a Redon drain was placed. The wound was sutured, and a sterile dressing was applied.

The surgery was challenging due to the fact that the ulnar artery, which was the only artery supplying the patient's hand, was located near the median nerve.

The patient underwent surgical release of the carpal tunnel in January 2023. The surgery was uneventful, and the patient reported significant improvement in her symptoms post-surgery. She reported no significant pain after surgery, but did experience some pulsating sensations in her fingers. No immobilization was applied. Patient was discharged with instructions for hand therapy and to avoid repetitive hand motions for a few weeks. At the follow-up appointment, the patient

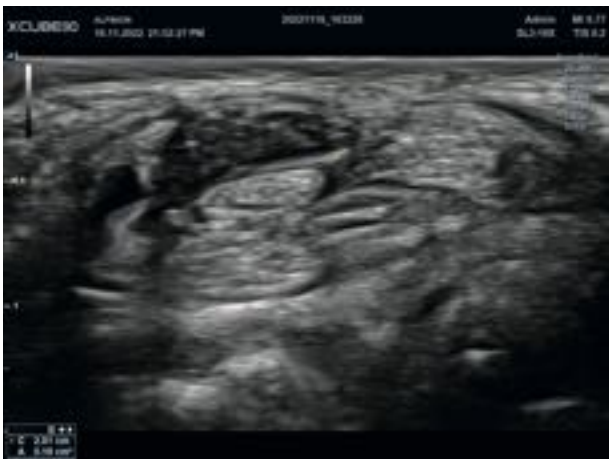

Figure 2. Transverse section through the median nerve proximal to the carpal tunnel. The cross-sectional area is enlarged, measuring 0.19 cm², which exceeds the normal range of less than 0.1 cm². The hypoechogenic ulnar artery is visible adjacent to the nerve

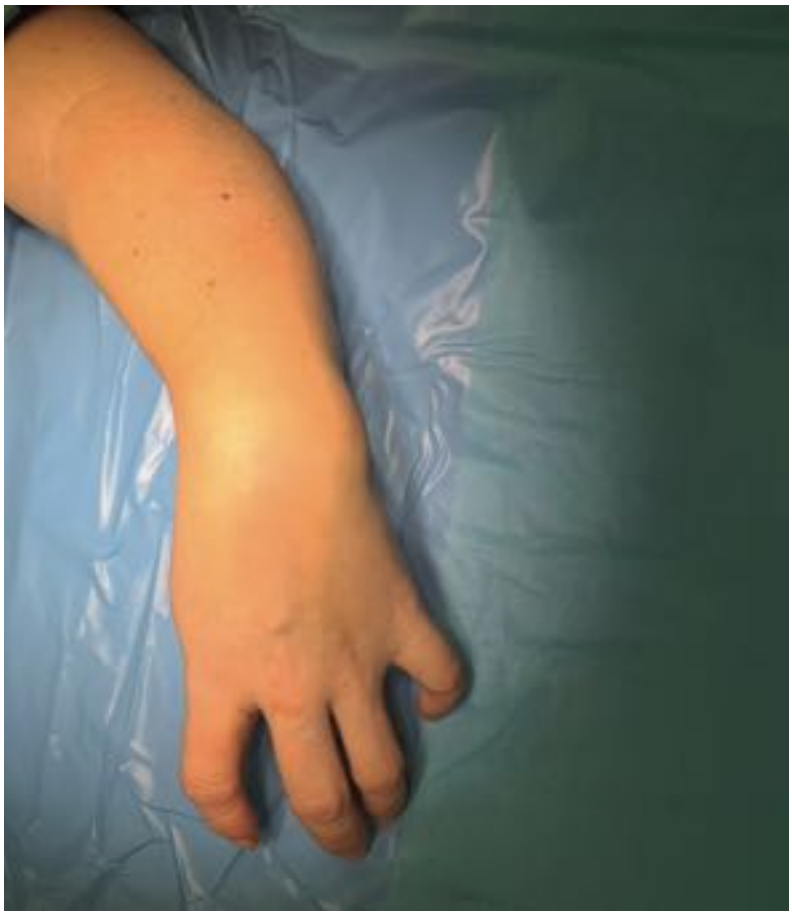

Figure 3. A patient's left hand prior to surgery

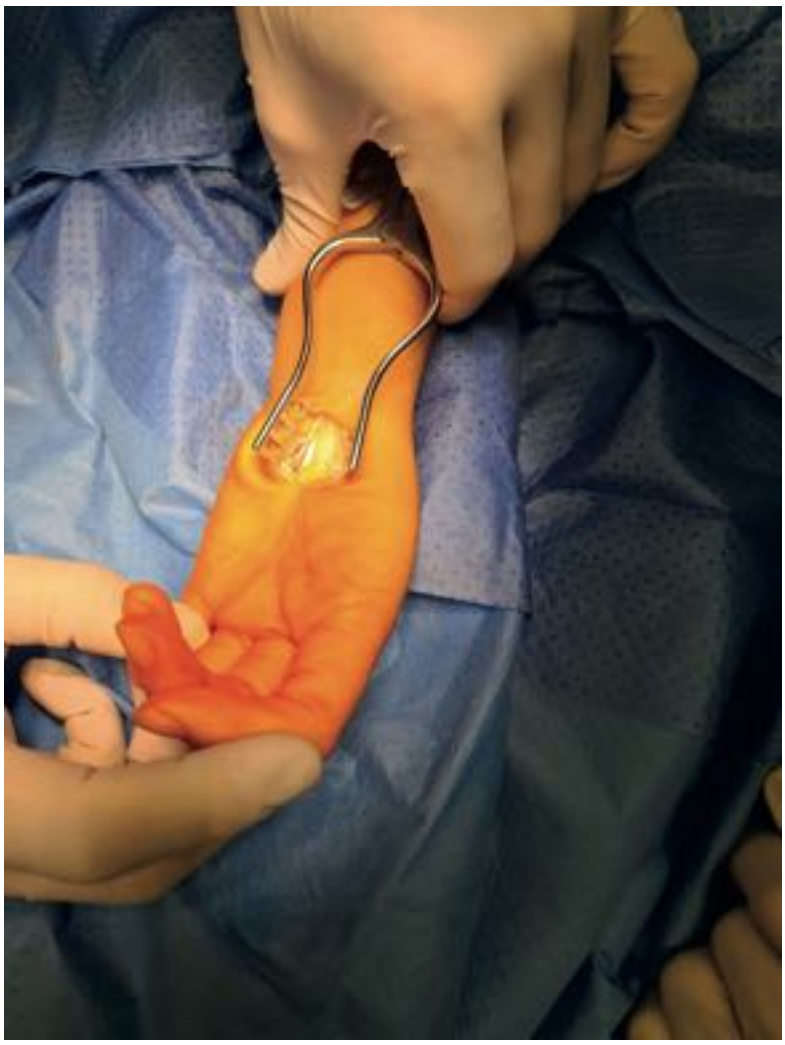

Figure 4. A narrowed nerve visualized after transecting the transverse ligament

reported full recovery of her hand strength and sensation. She had no complaints related to the surgery.

Discussion

The presented case report describes a patient with CTS, which is a common peripheral nerve entrapment disorder. The patient reported experiencing numbness, tingling, and weakness in her hand, which progressively worsened over several months, making daily activities such as holding objects and grooming difficult. The diagnosis of CTS was confirmed through ultrasound imaging, and the patient subsequently underwent surgical intervention to relieve the compression of the median nerve in the wrist.

Surgical intervention involving the transection of the transverse carpal ligament is considered the most effective treatment for releasing the nerve. The surgical decompression can be performed through different approaches, including the traditional open technique (involving a long longitudinal wrist incision and direct visualization of the ligament), minimally invasive approach (with a shorter wrist incision), or endoscopic technique (Padua *et al.*, 2016). In our practice,

we often opt for minimally invasive techniques, which have shown better outcomes compared to the standard open approach. These include fewer complications, higher patient satisfaction, improved symptoms, positive results on Tinel's, Phalen's, and compression tests, enhanced electrodiagnostic assessment, grasp strength evaluation, and faster recovery of the ability to perform personal tasks (Aslani *et al.*, 2012; Tarallo *et al.*, 2014; Elsharif *et al.*, 2014). However, in the case of the discussed patient, we decided to perform a longer incision to ensure better visualization of the ulnar artery. The use of Esmarch's tourniquet is a common technique for controlling ischemia during surgery. The insertion of the grooved probe under the ligament is a technique that can help protect the nerve during the procedure. The post-operative care of the patient will include monitoring for signs of nerve function recovery and wound healing. The use of a Redon drain and a sterile dressing is standard practice to prevent infection and promote healing.

It is worth noting the anatomical variations in arterial vascularization within the wrist when discussing this case. Anatomical variability of arterial vessels of the wrist is especially relevant in surgical procedures, such as wrist arthroscopy, which require a thorough understanding of the arterial anatomy of the region to avoid inadvertent injury to the vascular structures. The superficial palmar arch can be classified into three main types (Adachi and Hasebe 1928). The most common type is the ulnar type (typus ulnaris; 60%), which lacks a connection between the ulnar artery and the radial branch of the superficial palmar arch, which is then very weak. In the ulnar type, the area supplied by the ulnar artery may include, although rarely, all fingers. The next most numerous type is the radioulnar type (typus radioulnaris; 32%), in which both the ulnar artery and the radial branch of the superficial palmar arch supply the fingers. The least numerous type is the median-ulnar type (typus medianoulnaris; 8%), which occurs when the accompanying artery of the median nerve is well developed and replaces the branch of the radial artery; together with the ulnar artery, it then supplies the fingers. This type is also highly variable. The two vessels can join together to form a complete arterial arch, or they can branch directly to the fingers without connecting to each other. The median nerve's accompanying artery usually becomes the second finger's common digital artery. The radial half of the hand is mainly supplied by the deep palmar arch and dorsal metacarpal arteries, while the ulnar half mainly originates from the brachial artery. The position of the deep arch relative to the ulnar nerve branch is variable and if one palmar arch is diminished, the dorsal metacarpal arteries can compensate (Bochenek and Reicher 2012). The patient's case does not fit into any of the aforementioned three variants. The only artery observed in the patient at the wrist level was the ulnar artery. Furthermore, it was located very close to the median nerve, which posed a challenge during the surgery. Preoperative ultrasound examination of this finding enabled the surgeon to be prepared for this situation, avoiding any unfavorable surprises during the procedure.

The patient's positive treatment outcomes highlight the importance of early diagnosis and prompt treatment of CTS, as delaying treatment can result in further nerve damage and decreased quality of life. It also emphasizes the significance of considering a patient's medical history and potential risk factors in the diagnostic process.

There is currently no clear evidence to suggest that individuals with oligodactyly are at an increased risk of developing CTS. Nevertheless, certain sources appear to support the notion of a genetic predisposition to CTS (Hakim *et al.*, 2002). We have identified another case of carpal tunnel syndrome in an individual with thumb hypoplasia (Mace *et al.*, 2014). In this instance, the hypoplasia was part of Holt-Oram syndrome. The median nerve was significantly displaced towards the radial side, and a correlation

was suggested between this finding and the concurrent hypoplasia of the scaphoid bone. In a separate case (Robati *et al.,* 2009), a patient diagnosed with both carpal tunnel syndrome and Holt-Oram syndrome exhibited an unusual presence of the flexor digitorum superficialis muscle belly within the carpal tunnel, potentially contributing to the development of carpal tunnel syndrome.

Summary

Carpal tunnel syndrome is a common condition that can significantly affect a patient's daily life. Its diagnosis is typically straightforward, and treatment options range from conservative measures to surgical intervention. However, it is essential to consider a patient's medical history and perform examination and imaging to ensure correct diagnosis and appropriate treatment. In this case, the ultrasound examination in the patient with oligodactyly showed typical CTS image, but additionally we were able to find the absence of the radial artery which had an impact on the surgical procedure and increased our awareness during surgery.

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Authors disclose all financial and personal relationships that could influence their work.