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Case Report

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Vaughan Jackson Syndrome in Uncontrolled Rheumatoid Arthritis: RA Patient with Sudden Loss of Use with the Hands

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1. Abstract

Vaughan Jackson Syndrome is described as a disruption of the digital extensor tendons, beginning on the ulnar side of the hand and wrist with the Extensor Digiti Minimi and Extensor Digitorum Communis (EDC) tendon of the small finger. If the underlying pathology is not treated, sequential rupture of the EDC tendons of the ring, long, and index fingers occurs; rupture of the Extensor Indicis Proprius may follow. CASE REPORT: This is a case study of a 49 y/o female given treatment for 15 years. Lately, the patient has become tolerant leading to a dose escalation. Recently, she was presented with acute onset pain with limited motion of her hands. An MRI of each hand revealed a split interstitial tear of the Extensor Digitorum Minimi tendon along the fifth metacarpal diaphysis and extensive stenosing tenosynovitis around the right-hand Extensor Digitorum and Digiti Minimi tendons. These symptoms precisely reflect Vaughan Jackson syndrome, which was the primary diagnosis. With surgery normal function ability returned in each hand and wrist. DISCUSSION: It is important to recognize; the patient represents an unusual presentation of a common disease. However, the possibility of Vaughan Jackson Syndrome masquerading as Radial Nerve Palsy in uncontrolled RA should not be overlooked when treating an RA patient with severe disease and poor prognostic indicators including high rheumatoid factor with nodules.

2. Introduction

Rheumatoid Arthritis (RA) is a systemic multisystem inflammatory disorder with a predilection for small joint arthritis in a symmetric pattern [1]. Poor prognostic indicators include high titers of rheumatoid factor [2], and subcutaneous nodules of the skin [3] or lung [4], pleurisy [5], sicca [6], rheumatoid factor positivity [7,8], vasculitis [9,10] and other extra articular manifestations [11]. Vaughan Jackson Syndrome is described as a disruption of the digital extensor tendons, beginning on the ulnar side of the hand and wrist with the Extensor Digiti Minimi and Extensor Digitorum Communis (EDC) tendon of the small finger. If the underlying pathology is not treated, sequential rupture of the EDC tendons of the ring, long, and index fingers occurs; rupture of the Extensor Indicis Proprius may follow [12]. Figure 1 is a photo of a common presentation of Vaughan Jackson Syndrome [13].

RA is the most common underlying etiology of tendon rupture in the hand and wrist and is the usual clinical setting in which the term Vaughan-Jackson syndrome is employed [14].

Vaughn-Jackson Syndrome is a specific pathophysiologic process that leads to extensor tendon rupture in a rheumatoid wrist. While uncommon due to the introduction of disease-modifying anti-rheumatic drugs, early identification and referral to an orthopedic surgeon is vital to patient outcomes [14]. Other, less common patterns of tendon ruptures of the hands seen in RA should be ruled out,

including: 1) rupture of the extensor pollicis longus (EPL) in the vicinity of Lister's tubercle within the third dorsal compartment [15]; 2) Kienböck's disease, a condition of osteonecrosis of the lunate bone in the hand, and most patients present with a painful and sometimes swollen wrist with a limited range of motion in the affected wrist [16]; and 3) Mannerfelt syndrome, ruptures of the flexor pollicis longus and index-finger flexor digitorum profundus tendons within the carpal tunnel [17]. This is a case study of an RA patient showing symptoms leading to a clinical diagnosis of radial nerve palsy as part of mononeuritis multiplex actually having Vaughan Jackson syndrome. This disease can be treated surgically along with an early diagnosis and a prompt treatment [18]. An institutional review waiver was received.

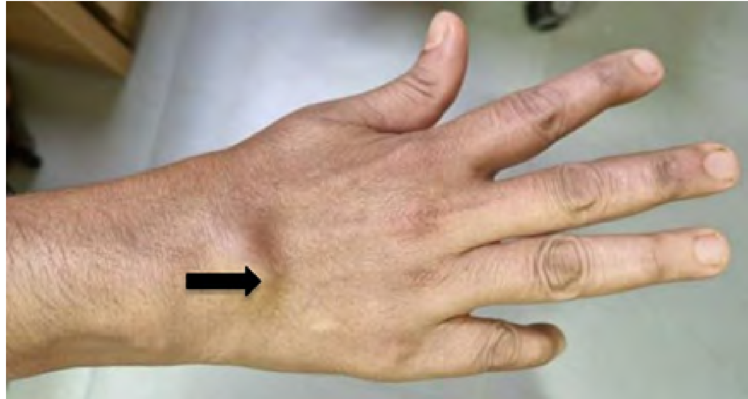

Figure 1: A photo of the dorsal hand and wrist of a patient with a common presentation of Vaughan Jackson Syndrome [13]. The arrow points to the area of spontaneous tendon rupture.

3. Case Study: Method and Results

The patient is a 49-year-old Vietnamese female with seropositive RA. Her rheumatoid factor is 2560 iu/ml (Normal <10 iu/ml), Cyclic Citrullinated Peptide (CCP) antibody is greater than 250 u/ml with severe disease. Given treatment for 15 years with methotrexate 20 mg/kg weekly, daily folic acid 1 mg, and infliximab starting dose 3 mg/kg Q8W, the patient had low disease activity. Over the past few years, the patient had become tolerant leading to a dose escalation being titrated to 10 mg/kg every 4 weeks with the addition of methylprednisolone, 4 mg. She remained stable at this dose. Subsequently her methotrexate was increased 25mg/week. We discussed switching biologic therapy however, due to fears on her part the switch to another Tumor Necrosis Factor (TNF) inhibitor was never tried. Recommendations included: B cell depleter, Cytotoxic T-lymphocyte antigen 4 (CTLA-4) inhibitor, and interleukin-1 and interleukin-6 inhibitors. The patient had gastroesophageal reflux disease (GERD) and was never given non-steroidal anti-inflammatory drugs (NSAIDs). X-rays of her hands were taken (Figure 2).

At the next follow up appointment, she presented with acute onset pain with limited motion of her hands. The patient is a right-hand dominant patient and noted the inability to open either hand. The patient experienced pain, weakness and swelling in both hands and wrists. She had subcutaneous nodules and pulmonary nodules, and always a high titer rheumatoid factor.

She is anemic with a hemoglobin of 10 grams. An MRI of each hand revealed a split interstitial tear of the Extensor Digitorum Minimi tendon along the fifth metacarpal diaphysis and extensive stenosing tenosynovitis around the right-hand Extensor Digitorum and Digiti Minimi tendons. These symptoms precisely reflect Vaughan Jackson syndrome, which was the primary diagnosis.

Orthopedics suggested surgical intervention to resolve the ongoing problem. They chose to operate on the dominant hand first then followed with surgery on the nondominant hand. The patient continued treatment with Prednisolone 10mg QAM (TNF inhibitor), underwent surgery with a bilateral stepwise approach, and did well with normal function ability in each hand and wrist allowing her to return to work as a casino dealer.

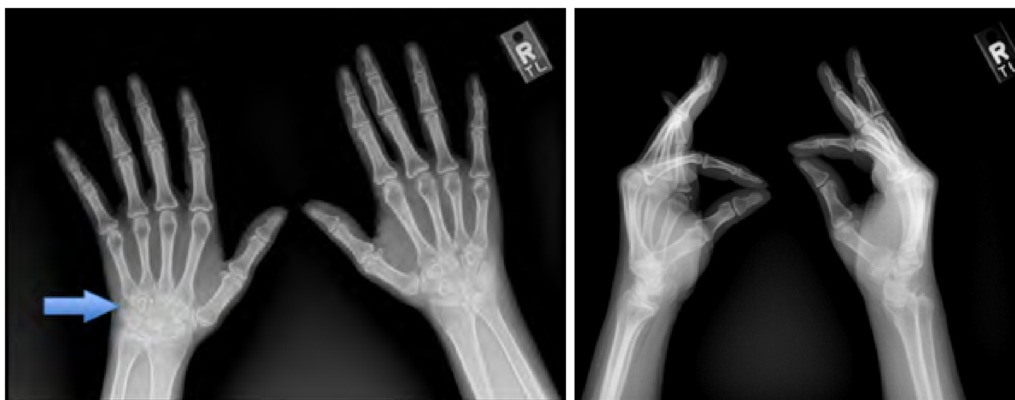

Figure 2: Anterior-Posterior (top) and lateral (bottom) views Kf the hands of the patient diagnosed with Vaughan Jackson Syndrome in Uncontrolled Rheumatoid Arthritis. The arrow points to an example of carpal coalition as seen in chronic Rheumatoid Arthritis.

4. Discussion and Conclusion

It is important to recognize; the patient represents an unusual presentation of a common disease. However, the possibility of Vaughan Jackson Syndrome masquerading as Radial Nerve Palsy in uncontrolled RA should not be overlooked when treating an RA patient with severe disease and poor prognostic indicators including high rheumatoid factor with nodules.

Fortunately, this patient did not have interstitial lung disease nor vasculitis (significant characteristics of uncontrolled RA) but did have acute tendon rupture involving multiple tendons and aponeurosis. Prompt recognition of the condition and referral to a hand surgeon left her in stable condition and able to continue employment.

In summary, the patient presented with a lack of extension in the dorsum of the hand, and originally was felt to have flexion, for which wrist drop was considered due to indications of radial nerve disease or mono neuritis multiplex. Therapy using pulse steroids and Cytoxan was considered. However, due to the distribution of various fingers having different degrees of strength, an MRI was ordered. MRI findings included extensive stenosing tenosynovitis around the Extensor Digiti Minimi tendons and a split interstitial tear of the right Extensor Digiti Minimi tendon along the fifth metacarpal diathesis. Similar findings were found in the left wrist. With continued therapy, as described above, she has returned to work and is doing well.

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