Case Report:
Carpal Tunnel Syndrome Associated with Klippel-Trénaunay Syndrome: A Case Report

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Background and Importance: Klippel-Trénaunay syndrome is a rare congenital condition that rarely affects peripheral nerves. Median nerve involvement at the carpal tunnel level has only been reported on four occasions of this syndrome in the medical literature.

Case Presentation: A 61-year-old Caucasian female patient with Klippel-Trénaunay syndrome presented with a 10-month history of paraesthesia and numbness affecting the median nerve distribution area of her left hand. The clinical and neurophysiological examination confirmed a moderately severe carpal tunnel syndrome that required surgical decompression.

Conclusion: Pre-operative findings demonstrated diffuse vascular infiltration and engorgement of the median nerve. Flexor tendons had a normal appearance. The patient had a satisfactory post-operative period with full resolution of her symptoms 4 weeks after the procedure. Although peripheral nerves are rarely affected in patients with Klippel-Trénaunay syndrome, neurological symptoms could indicate nerve involvement and Magnetic Resonance Imaging (MRI) examination should be considered to further assess the extension of the lesion.

ABSTRACT

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Keywords:
Klippel-Trénaunay syndrome, Carpal tunnel syndrome, Median nerve

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1. Background and Importance

Klippel-Trénaunay syndrome is an uncommon congenital condition that can occasionally affect peripheral nerves [1]. In the medical literature, the median nerve has only been found to be affected in 4 cases [2-5], but despite this, we believe that sonographic or MRI imaging of the carpal tunnel and the median nerve should be considered pre-operatively.

2. Case presentation

We present the case of a 61-year-old patient with Klippel-Trénaunay syndrome complaining of a 10-month history of paresthesia affecting the median nerve distribution of her left hand. The clinical and neurophysiological examination confirmed the diagnosis of carpal tunnel syndrome and the patient was operated on, with full resolution of her symptoms.

On examination, the patient presented the classical port-wine stains distributed on the entire left upper extremity skin. At hand level, she had a normal abductor pollicis brevis muscle with positive Tinel, Durkan, and Phalen tests. Neurophysiological investigations confirmed a moderate reduction of the median nerve conduction velocities across the left wrist.

The patient was taken to the theater for carpal tunnel release under local anesthesia. Inspection of the median nerve revealed a diffuse vascular infiltration and ingurgitation with an enlarged diameter of the nerve and indentation with hourglass deformity (Figure 1). Flexor tendons had a normal appearance. The immediate post-operative period was uneventful and two weeks later, the patient was reviewed for suture removal and gradual mobilization. At that time, most neurological symptoms had already improved and the patient was discharged 4 weeks after the operation with full resolution of symptoms with an open appointment.

Figure 1. Median nerve showing diffuse vascular infiltration and ingurgitation with enlarged diameter and indentation with hourglass deformity
3. Discussion

Klippel-Trénaunay syndrome is a rare congenital condition characterized by hypertrophy of bone and soft tissue, varicose veins, and cutaneous capillary malformation developing the classical skin port-wine stains [1]. Carpal tunnel syndrome is seldom associated with Klippel-Trénaunay syndrome and only 4 cases (Table 1) have been reported in the medical literature [2-5]. This is explained by the fact that peripheral neuropathy is rare in Klippel-Trénaunay syndrome. It is related to the presence of epineurial microscopic arteriovenous anastomoses and endoneurial vascular coils which corresponds to the macroscopic findings of the case presented in this article.

Lymphatic obstruction, fibrolipomatous hamartoma, and cavernous hemangioma have been described as pathogenesis in this type of neuropathy. Furthermore, the risk of malignancy of these vascular nerve lesions has been described [6].

Despite its rare association with peripheral neuropathy, we believe that patients with Klippel-Trénaunay syndrome who complain of persistent paresthesia in a specific nerve distribution would benefit from a further diagnostic tool such as MRI to fully assess the extent of the disease. A Tinel test would be relevant to determine the site of imaging of the relevant nerve.

4. Conclusion

Patients with Klippel-Trénaunay syndrome who complain of neurological symptoms, apart from compressive neuropathies, should be assessed for nerve involvement secondary to their congenital condition. Clinical examination including the Tinel test and neurophysiology studies together with imaging techniques such as MRI will give the physician and surgeon the full picture of the condition. Nerve infiltration with epineurial arteriovenous anastomoses and endoneurial vascular coils will inevitably cause an enlarged nerve diameter that can lead eventually to nerve compression in sites where this is anatomically feasible.

Ethical Considerations

Compliance with ethical guidelines

Pre-operative written informed consent was obtained from the patient to take the images.

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Conflict of interest

The author declared no conflicts of interest.

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References


