Pronator Syndrome Due to Schwannoma

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Abstract This report presents a rare case of pronator syndrome due to median nerve compression by Schwannoma. The tumour was derived from the median nerve and was located between the two heads of origin of the pronator teres muscle in the left antecubital fossa. The patient was completely cured after excision of the tumour.

Keywords Compression neuropathy · Median nerve · Pronator syndrome · Neurilemmoma · Schwannoma

Introduction

Seyffrath is credited for the first description of pronator syndrome [1]. Pronator syndrome is defined as the compression of the median nerve at the proximal forearm at one or more of the four following structures: an anomalous Struther ligament, the lacertus fibrosus in the antecubital fossa, two heads of origin of the pronator teres muscle and the proximal arch of the flexor digitorum superficialis muscle. However, proximal compression of the median nerve may occur by abnormal anatomical structures around the elbow joint [2–5]. The median nerve may be also compressed by space occupying lesions in the antecubital region [6–9]. Schwannoma or neurilemmoma is a benign tumour of the myelinating Schwann cells of the nerve sheath [10–12]. This report describes a rare case of pronator syndrome due to Schwannoma of the median nerve in the antecubital region.

Case Report

A 30-year-old man was referred to our clinic because of a sensory disturbance in the thumb, index and middle fingers as well as the radial half of the ring finger of his left hand that had persisted for over 1 year. Seven months prior to referral, he noticed swelling in his left antecubital fossa. Since then, his symptoms progressively increased and pain and weakness in the forearm became more apparent over the course of 3 months. His symptoms were exacerbated by his occupational activities. He had no significant medical or surgical history. Laboratory test results were normal. Upon palpation of antecubital fossa, there was a hard mass lateral to the proximal flexor muscle mass. The patient's symptoms were intensified by resistive tests of elbow flexion and pronation of the forearm against resistance. Power grip was 40 kg. (90 lbs.) in the left hand and 55 kg. (120 lbs.) in the right hand. No individual muscle weakness was noted. Tinel's sign was positive upon percussion of the median nerve in the antecubital fossa. Carpal tunnel syndrome was ruled out because Tinel's sign, Phalen's and Durkan's tests were negative at the wrist. Cervical radiculopathy and thoracic outlet syndrome were also ruled out because Spurling's and Adson's signs were absent. A working diagnosis of pronator syndrome due to median nerve compression by a tumour was made.

A nerve conduction study demonstrated a partial conduction block at the level of the proximal forearm; however, electromyography was normal. Plain radiography of the left elbow was normal. Ultrasound examination revealed a lobulated soft tissue tumour with a cystic component, adjacent to the median nerve in the proximal forearm. This finding aroused the suspicion that the tumour may have a nerve sheath in origin, and the cystic appearance on ultrasound was in favour of Schwannoma; however, the major differential diagnosis included was ganglion cyst.

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Magnetic resonance imaging (MRI) sagittal view demonstrated a hyper-intense tumour in the antecubital fossa (Fig. 1). MRI coronal view demonstrated a hyper-intense tumour adjacent to the neurovascular structure in the antecubital fossa (Fig. 2). MRI axial view demonstrated a hyper-intense tumour that was surrounded medially by the flexor muscle mass in the antecubital fossa (Fig. 3).

At surgery, the bicipital aponeurosis (lacertus fibrosus) was released. The tumour was found between the two heads of origin of the pronator teres muscle, adjacent to the median nerve. The tumour was found to be an eccentric bulging of the median nerve (Fig. 4) which was well-encapsulated mass measuring $5 \times 3 \times 3$ cm (Fig. 5). Under loop magnification, the tumour was dissected away from the median nerve fascicles.

Histopathology showed an encapsulated tumour composed of spindle shaped cells with oval to elongated normochromic nuclei. The nuclei were arranged in a fascicle and whorl pattern. There was no Verocay body (Fig. 6). Furthermore, upon histopathological examination, hypocellular myxoid areas, aggregation of foamy macrophage, cystic degeneration, thick-walled hyalinized blood vessels with congestion and thrombus formation, hemorrhage and infiltration of inflammatory cells were observed. Immunohistochemistry staining with S-100 was positive. These findings were consistent with a diagnosis of benign Schwannoma (Fig. 7).

Fig. 1 MRI sagittal view demonstrates hyperintense tumour in the cubital fossa. (Arrows indicate the tumour. The star indicates the distal humerus)

Fig. 2 MRI coronal view demonstrates hyperintense tumour adjacent to neurovascular structure in the cubital fossa. (Large arrows indicate the tumour. Small arrows indicate the neurovascular)

Fig. 3 MRI axial view demonstrates hyperintense tumour located between the flexor muscles mass in the cubital fossa. (The stars indicate the tumour. Small arrows indicate the neurovascular)

Fig. 4 At surgery: the tumour was derived from the median nerve. (The *star* indicates the tumour and the *arrow* indicates the median nerve)

There was no median nerve deficit after surgery. The patient's symptoms improved after surgery and he was completely cured after 6 months.

Discussion

Schwannoma or neurilemmoma is a benign tumour of the myelinating Schwann cells of the nerve sheath [10–13]. Schwannoma is the most common tumour of the peripheral nerves [7]. Schwannomas develop along the nerves, so they are transversely mobile but not longitudinally mobile. Macroscopically, Schwannoma is a well capsulated tumour that produces an eccentric bulging of the involved nerve. Schwannoma can be removed without injury to the nerve fascicles. Microscopically, Schwannoma is characterized by two types of cellular arrangement: Antoni A and Antoni B. The Antoni A pattern consists of compact spindle cells arranged in bundles and cords. The Antoni A cells may be arranged in palisade rows that are well known as Verocay bodies. The Antoni B pattern consists of scattered and irregular spindle cells separated by loose myxoid stroma. Usually

Fig. 5 The tumour with a nodular and firm consistency on palpation measuring 5×3×3 cm

Fig. 6 Tumour composed of spindle shaped cells with oval to elongated normochromic nuclei. The nuclei were arranged in fascicle and whorl pattern. (Hematoxyline and eosin staining×400)

Schwannoma contains both cellular arrangement types; however, either Antoni A or Antoni B arrangement may predominate. Immunoreactivity on S-100 immunohistochemistry staining confirmed that the origin of the spindle cells was the Schwann cells. Malignant transformation of Schwannomas is a rare occurrence [6–8, 10–13].

In the current case, ultrasound examination showed that the tumour was adjacent to the median nerve. Cystic appearance on ultrasound was in favour of Schwannoma [14]. MRI is very helpful for diagnosis of nerve sheath tumours. The 'target sign' on MRI is characteristic for Schwannoma and neurofibromas. The 'target sign' is an image with central low signal intensity and peripheral high signal intensity on T2-weighted images. This sign may be seen in 15–54 % of Schwannomas and 50–70 % of neurofibromas [10, 11]. In the current case, MRI did not demonstrate the typical 'target sign', but MRI contributed to the diagnosis by delineating the anatomical relationship of the tumour and the surrounding muscles and neurovascular structures.

Fig. 7 Immunohistochemistry staining with S-100 is positive

The diagnosis of pronator syndrome is usually difficult [2–5]. Its symptoms overlap with those of the carpal tunnel syndrome, which is more frequently diagnosed. Clinical findings of pronator syndrome are inconsistent. The value of the described provocative maneuvers to evaluate the subjective symptoms of patients is unclear [2]. Tinel's sign is not necessarily observed. Only 10 % patients with pronator syndrome may have abnormal electrodiagnostic findings [2–5]. In the current case, presence of a space-occupying lesion in the proximal forearm adjacent to the median nerve, electrodiagnostic findings, patient's symptoms and MRI findings confirmed the pronator syndrome diagnosis. Double crush syndrome was ruled out because the signs were absent. Pronator syndrome caused by Schwannoma is a rare occurrence. Schwannomas around the elbow joint have rarely been reported [6–8, 11, 13]. Haussmann has reported a case of pronator syndrome caused by malignant Schwannoma of the median nerve [6]. The current report adds another rare case of pronator syndrome caused by median nerve Schwannoma to the academic literature.

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