CASE REPORT

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Schwannoma of the Ulnar Nerve in the Elbow: A Case Report

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Schwannomas are soft tissue sarcomas arising from neurilemma of Schwann cells in peripheral nerves, and is the most frequent type of benign tumor found in these nerves. We report a case of a Schwannoma of the ulnar nerve in the elbow joint, and along this report, give a review of the literature. A 46-year-old male patient was hospitalized with complaints of swelling and pain in the left elbow and a tingling sensation and hypoesthesia of the fourth and fifth fingers. Physical examination of the patient showed he was positive for Tinel's sign, and magnetic resonance imaging results demonstrated the presence of a Schwannoma. Subsequent biopsy and excision of the Schwannoma was carried out. The suspected mass, which had a clear margin separating it from the healthy nerve of the medial left elbow, was removed along with its 2 × 2 × 3 cm capsule after a histological diagnosis of a Schwannoma was made. Pathophysiological results confirmed the excised mass as a Schwannoma. Schwannoma of the ulnar nerve within the elbow joint is rare and differential diagnosis is difficult. Therefore, treatment can only proceed after the presence of Schwannoma has been confirmed by physical and radiological examinations.
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Key Words: Elbow joint; Ulnar nerve; Neurilemmoma; Schwannoma

Schwannomas are soft tissue sarcomas arising from neurilemma of Schwann cells in peripheral nerves and comprise 5% of all soft tissue sarcomas. Although they are the most frequent type of benign tumor found in peripheral nerves, these benign Schwannomas rarely transform into malignant tumors.[1] The proportion of Schwannomas that occur in the upper extremity is 0.8−2.1% of all tumors, whereas this value increases to 19% when the wrist region is included in the calculation. This means that Schwannomas of the upper extremity generally occur in the hand or wrist regions, whilst it does not generally occur in elbows or the upper arms.[2] However, recent literature by Tang et al.[3] has reported of an occurrence of up to 75% of Schwannomas in the elbow and the distal region. We report a case of a Schwannoma of the ulnar nerve in the elbow joint, and along this report, give a review of the literature.

Case Report

A 46-year-old male had received conservative management for a recurrent swelling in the lateral elbow over a period of 2 years. From last year, however, the patient complained of additional, persistent symptoms of pain in the elbow, tingling sensation and hypoesthesia of the 4th and 5th fingers, which we found was the result of a mass at the medial elbow joint. Physical examination of the elbow indeed showed a soft, palpable, oval-shaped mass with a 2 × 3 cm dimension at this region. A positive Tinel's sign was seen as radiating pain and tingling sensation along the forearm ulnar when the medial elbow was tapped. Although plain radiographs of the transverse, lateral, and planar planes of the elbow did not show anything unusual, magnetic resonance imaging (MRI) results showed a 2 × 2 × 3 cm-sized mass at the Cubital tunnel and a low-signal, whose signal intensity was comparable to that of its surrounding muscle, within the medial elbow joint in a T1-weighted image (Fig. 1A).

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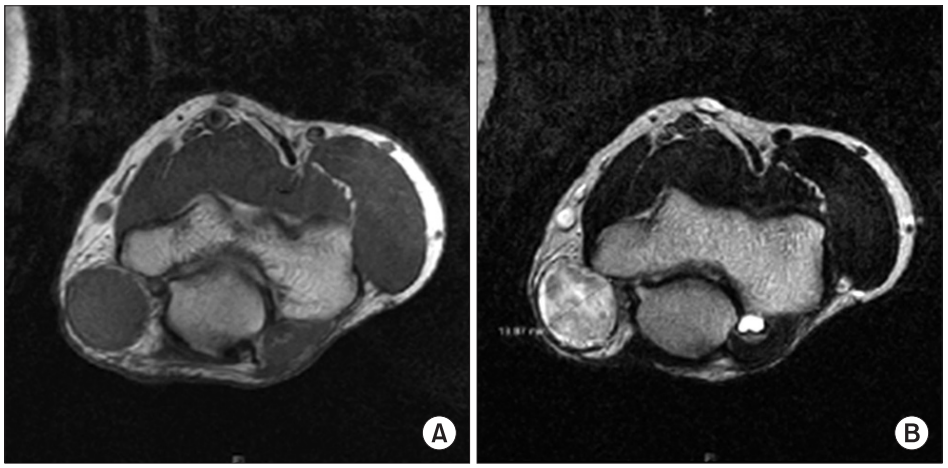

Fig. 1. (A) Axial T1-weighted image showed ovoid mass with low signal intensity which is isointense to surrounding muscle. (B) Axial T2-weighted image showed ovoid mass with high signal intensity. Compose of hypointensity on marginal area and hyperintensity on central area.

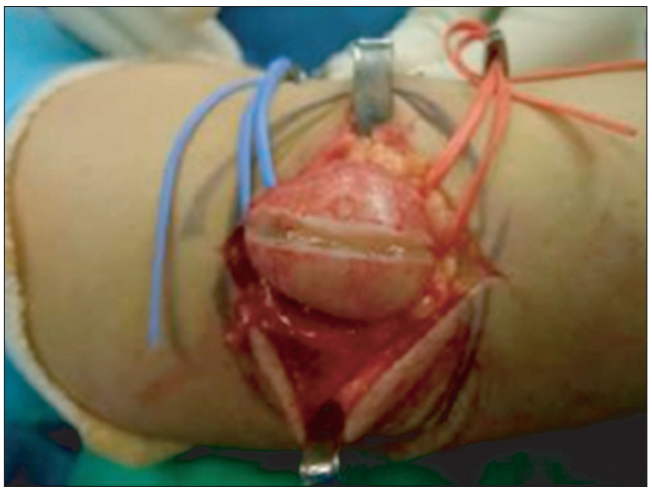

Fig. 2. An ovoid mass of white-yellow soft tissue, measuring 3 × 2 × 2 cm (intraoperative picture).

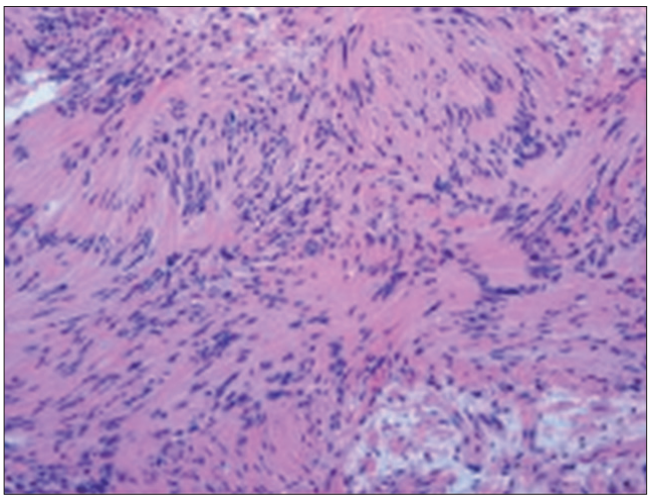

Fig. 3. Microscopic section shows spindle cells in palisading pattern, Antoni A pattern (hypercellular region) and Antoni B pattern (hypocellular region) (H&E, ×200).

In comparison, T2-weighted images showed a high-intensity signal with slight heterogeneity (Fig. 1B). Intra-mass hemorrhage and septum formation were also noted. To begin treatment and for histological examination, biopsy and excision of the mass were performed under general anesthesia. The surgery was performed with the upper body rotated laterally and in supination so that the posteromedial elbow joint was in good view. A 7 cm-skin incision was made from the anterior to the distal side of the mass. Further, to locate the root of the flexor forearm, a transverse incision was made, and then the skin was pulled anteriorly and fixed. Next, the ulnar nerve penetrating through the posterior side of the medial mass was exposed.

We found a soft brown mass of 2.0 × 2.5 cm dimension covered by a white capsule. A complete excision of the mass was possible as it was clearly distinguishable from the surrounding unaffected nerves and soft tissue (Fig. 2). A neat removal of the mass allowed us to put enough pressure on the ulnar nerve, to wash the area of surgical intervention, and to suture the incision. When the mass was excised, we found partial bleeding at the center of the mass, and that it was filled with light yellowish tissue. Histologically, the mass showed a palisading pattern with a mixture of smooth Antoni A and rough Antoni B-type cells that are characteristic of Schwannomas, thus enabling us to make a diagnosis (Fig. 3).

Discussion

Schwannomas are benign tumors that originate from Schwann's cell and comprise 5% of all soft tissue sarcomas. Most Schwannomas are covered by a neural capsule and rarely transform into malignant tumors.[1] There is no gender bias in the contraction of Schwannomas and prevalence across ages 20 to 60 is fairly even, except during middle-age, during which you are more likely to contract Schwannomas.[2] Schwannomas become larger over time but rarely exceed a diameter of 3 cm,[2] which

is why in most cases Schwannomas gradually put pressure on neural bundles, delaying visible symptoms of Schwannomas and thus diagnosis until the late stage of tumorigenesis.[2] Fortunately, in most cases Schwannomas remain as a distinct mass from neural tissues allowing relatively easy and complete surgical removal of these masses without causing neural deficits.[4]

Our case report describes a patient who had recurrent symptoms for over 2 years, but was only diagnosed of Schwannoma later on. This was because a mass that was initially impalpable became palpable over time by physical examination. Subsequent diagnostic MRI detected the mass at the medial elbow joint after which appropriate surgical interventions could be implaced. The proportion of Schwannomas that occur in the head and neck is 45%, in the upper limbs is 19%, and in the lower limbs is 13.5%. Those that occur in the upper limbs tend to occur in large nerves such as the ulnar or the median nerves.[2] Within the upper limb, Schwannomas can also occur in nerves found in any region between the brachial plexus and the hands, but usually it is found in the volar than the dorsal region, and the Schwannomas found in these regions together make up 0.8−2.1% of all Schwannomas.[2]

As aforementioned, neural deficits caused by Schwannoma are rare, but neurological symptoms arising from mass-induced pressure on nerves have been reported, the symptoms of which is a positive Tinel's sign, pain around the area of pressurization upon gentle tapping.[2] Approximately 30−70% of Schwannoma patients experience such pain and over 20% complain of hypoethesia.[2] A recent report on a group of Schwannoma patients found all of them had to some degree hypoethesia and/or Tinel's sign.[3] The non-specific symptoms of Schwannomas mean that suspecting someone of a soft tissue sarcoma or neuroma is difficult just by physical examination or even by plain radiography and differentiating it from tenosynovitis or Cubital tunnel syndrome is challenging.[5,6] Even for such symptoms, a physician should consider a slowly growing independent tumor that may originate deep inside the nerve where palpation induces a transverse but not a verticakl movement of an otherwise fixed mass.[4] In this report, we found a palpable mass that grew at least over 2−3 years. The patient showed a positive Tinel's sign and hypoethesia at the fingertips following the ulnar nerve. As well as diagnosing the patient with a mass-induced Cubital tunnel syndrome, we were able to diagnose and subsequently surgically treat the patient with Schwannoma. In general, for its low cost and speed, ultrasound is used to make a differential diagnosis at the initial stage.[7] On ultrasound, the contour of the Schwannoma is clear and has a low, homogeneous signal.[7] Further, by following the length of the nerve and examining its continuity, it is possible to distinguish Schwannomas from neuromas of other soft tissue origin.[7] Höglund[7] suggested that ultrasound has 59% diagnostic accuracy for neuromas but it cannot be used with enough specificity to diagnose other types of soft tissue neuromas. Similarly, Fornage[8] showed that 50% of Schwannomas was detected using standard ultrasound findings, of which 75% of findings could be used to indicate where the contours of the tumors were exactly. As such, current literature shows that ultrasound cannot diagnose Schwannomas with a high level of accuracy, but its cost-effectiveness and speed makes it a useful first-line diagnostic tool.

Although the cost is greater than when making a diagnosis using ultrasound, MRI can determine the position and origin of tumors, detect any associated neurovascular cells, and thereby be a better first-line of approach to gain information for surgical intervention.[6] Characteristically, Schwannomas appear as a radial signal in T1-weighted images and is found in the orientation of the nerve as a heterogeneous, high-intensity signal.[1,7] Contrast-injected weighted imaging using Gadolinium does not show a heterogeneous signal.[1,7] Further, for Schwannomas, a characteristic split fat sign, which refers to fat tissue surrounding the neurovascular structures of the muscle, is detected in T1-weighted images, and a target sign, which is low-intensity signal at the centre of the tumor and a high intensity signal surrounding the tumor, is detected in T2-weighted images.[9] Histologically, the target sign would show an increase in signal intensity in the area surrounding the tumor and a decrease in signal at the centre of the tumor in a T2-weighted image when contrast medium is injected. This difference in change in signal intensity is because of the difference in the composition of cells. The surrounding cells are composed mostly of myxoma, whereas the centre contains mostly collagenous fiber.[9]

Degeneration can occur for relatively large Schwannomas, called ancient Schwannomas, which can be detected by the formation of a cystoma or a bleeding-induced septum at the Schwannoma cross-section.[6] Although imaging results of ancient and non-ancient Schwannomas are similar, the presence of a cystoma is indicative of an ancient Schwannoma. Cystomas can be detected as a low intensity signal in T1-weighted images and a high intensity signal in T2-weighted images, and the signal intensity remains unchanged even when contrast media is injected.[6] In rare incidences, signal intensity around the cystoma or the capsule may enhance as a result of contrast injection.[10] Although Schwannomas tend to be eccentric to the nerve, they cannot always be neatly separated and up to 30% of cases have associated neurofibromas, which may or may not be capsulated.[10] Thus, even though MRI is a relatively expensive procedure that cannot completely differentiate between Schwannomas and neurofibromas, it is most useful to use as a diagnostic tool and to confirm the position of the mass and associated neurovascular structures prior to surgical intervention.[9]

We performed diagnostic radiology using MRI. We found a low intensity signal in T1-weighted images, showing a signal comparable to signals given by muscle, and a low intensity signal centrally and a high intensity signal at the vicinity of the lesion,

deduced as target sign, in T2-weighted images. Thus, we were able to diagnose the patient with Schwannoma and prepare the following procedures using the results of the MRI.

In our case report we described a palpable mass, confirmed by physical and radiological examination, which was treated by surgical pressure. After surgery, the patient achieved relief of symptoms. However, for many Schwannomas a palpable mass cannot be seen. When patients who complain with neurological symptoms of the upper limb peripheral nerves are examined, it is often that rare neural tumors such as Schwannoma are overlooked even though thorough standard physical tests are made. Though rare, a Schwannoma or other types of neural ganglion cysts cannot be ruled out for patients who show no clear evidence of tumor after physical or radiological examinations, but show positive Tinel's sign, pain, and/or neural deficits. Differential diagnosis should include diseases such as Cubital tunnel syndrome. Although both ultrasound and MRI lack specificity, they can be used as first-line approaches to differentially diagnose Schwannomas with sufficient sensitivity. Early detection and intervention is warranted for Schwannomas as failure to do so can lead to prolongation of symptoms.

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