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Case Report

Golf ball near elbow: a rare presentation of a giant ulnar nerve Schwannoma

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ABSTRACT

Schwannoma is a type of peripheral nerve sheath tumor but can produce a mass effect with increase in size, spontaneous pain, paresthesia and motor weakness being the main complaints. A 60-year-old male patient presented with swelling in the left forearm since the past 8 months and numbness and tingling sensation in the hand for 1 month. Examination revealed a solitary lesion in the ulnar aspect of left proximal forearm with glove and stocking type of neuropathy, wasting of hypothenar eminence and amputation of left 5th digit, with hypopigmented lesions over the forearm and leg. Slit skin smear and biopsy of hypopigmented lesion was done to rule out leprosy. Ultrasonography (USG) and contrast-enhanced magnetic resonance imaging (CE-MRI) of left forearm lesion revealed homogenously enhancing lesion involving the proximal portion of left forearm - suggestive of peripheral nerve sheath tumor. Hence the patient was treated with excision and biopsy. Histopathological examination revealed a giant Schwannoma (6×4×3.5 cm) of the ulnar nerve with typical findings of spindle shaped cells few verocay bodies. Despite rare, tumors should be taken into account in the differential diagnosis in such presentations. It is important to remember that Schwannoma, in these cases, is the most common tumor. The simple removal of the tumor after careful dissection is generally enough since the recurrence and malignant transformation rates are low.

Keywords: Giant, Schwannoma, Ulnar nerve

INTRODUCTION

Primary neural tumors of the upper extremity are rare and represent less than 5% of soft-tissue neoplasms of the upper extremity. Among peripheral nerve tumors, neurilemmomas are the most common and are communally known as Schwannomas once they originate from the cells of the Schwan.

A schwannoma is usually a benign nerve sheath tumor composed of Schwann cells and is a type of nerve sheath tumor which normally produces the insulating myelin sheath covering peripheral nerves. It is most common type of benign tumor of peripheral nerves of upper limb.[1] They are non-invasive tumours arising from peripheral nerve sheaths and are encapsulated by epineurium. Most are asymptomatic, but can produce a mass effect with increase in size. Spontaneous pain or pain after working, paraesthesia and motor weakness are the main complaints. The World Health Organization classifies schwannoma as a grade I benign tumor.

About 90% of the schwannomas are sporadic and may have a possible genetic etiology, where NF2 gene on chromosome 22 has been suggested to play an essential role in their development.[2] Schwannomas are composed of two types of tissue known as Antoni A and B and the Tumor cells are highly immunopositively for S-100 protein. Based on the cellular components, they are mainly classified into: plexiform schwannoma, cellular schwannoma, and melanotic schwannoma.

Sites of involvement include the subcutaneous nerves of the face and neck, extremities and deep tissues of the

retroperitoneum and mediastinum. Usually, this tends to occur in patients aged 30–60 years and has no race or sex predisposition. They are usually slow growing and can remain symptom free for many years. The mean age for their diagnosis is about 56 years. Tenderness is felt when palpating the mass. Neurologic symptoms or paraesthesia due to pressure on the nerve may be present rarely if the tumour is large and symptoms can appear at the corresponding anatomical area.

Magnetic resonance imaging (MRI) and computed tomography (CT) scans are recommended, but MRI is preferred. They usually appear as oval or round mass with an isointense or hypointense signal on T1-weighted images and a hyperintense, heterogeneous signal on T2-weighted images giving rise to a target sign.[3,4]

While different techniques are present to manage schwannomas, small asymptomatic schwannomas are left for observation. Surgical resection and radiotherapy are used for progressive and/or symptomatic lesions.[1] The prognosis is excellent as recurrence after total resection is uncommon. Malignant changes may occur in long-standing schwannomas, although this is rarely reported.[5]

CASE REPORT

A 60-year-old male presented with swelling in the left forearm that had developed over the past eight months, accompanied by pain in the last one month. The swelling was insidious in onset, initially small, but progressively enlarged to approximately 3.5×5 cm in size. The patient reported dull, aching pain of moderate intensity, exacerbated by strenuous activities, lifting weights, and described numbness and a pricking sensation in the ulnar distribution of the left hand along the ulnar one third for the past one month.

The patient denied any history of loss of grip, recent trauma, or similar swellings elsewhere. Notably, there was a history of traumatic amputation of the left fifth digit. He had history of epilepsy for which he was receiving treatment with carbamazepine. No other significant comorbidities were reported.

General physical examination

Physical examination revealed multiple hypopigmented patches on the left upper limb and bilateral lower limbs. Amputated stump was noted in little finger of left hand and right great toe (Figure 1).

Inspection

The left forearm examination noted a solitary, firm, non-tender swelling located on the anteromedial aspect of the proximal third of the forearm, which was non-fluctuant, non-compressible, and fixed to the underlying structures but not to the skin. Swelling was around 3.5×5 cm in size, firm in consistency, non-tender, oval in shape with well-defined edges. It was non-fluctuant, non-compressible and non-reducible, non transluminiscent and no palpable pulse was present. It was Fixed to underlying surface but not to skin. Skin over the swelling was normal with no local rise of temperature.

There was glove-and-stocking type neuropathy in both upper limbs and evidence of muscle wasting in the hypothenar region of the left hand. Mobility was restricted along the longitudinal axis and no regional lymphadenopathy was noted. Pressure effect was positive and altered sensation was noted in the hand and proximal 1/3rd of left forearm. Sensation was altered in the left hand, with a positive Tinel's sign over the ulnar nerve distribution.

Differential diagnosis

The differential diagnoses included: neuro-lipoma, Hansen's disease, nerve sheath tumors (schwannoma, neurofibroma), and hematoma.

To rule out Hansen’s disease, slit skin smears and skin biopsy of hypopigmented areas (Fite-Farraco stain) were performed, both yielding negative results for lepra bacilli.

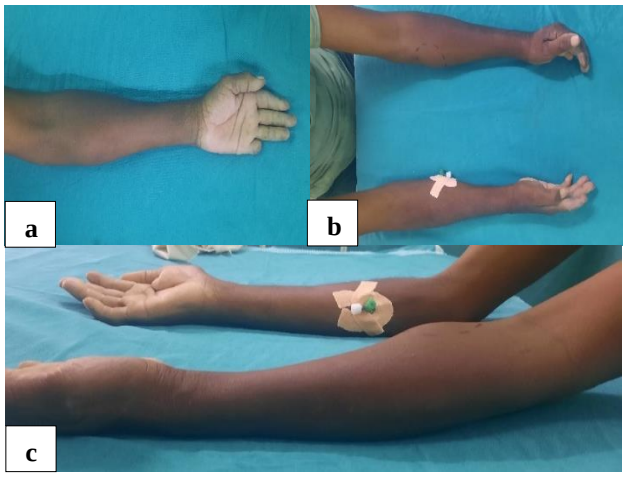

Figure 1: Swelling in anteromedial aspect of proximal 1/3rd left forearm.

Investigations

Pre-operative investigations included routine blood tests and imaging. MRI was suggested to evaluate the extent of the swelling and its relationship to surrounding structures, focusing on the characteristics typical of nerve sheath tumors. The MRI findings confirmed presence of peripheral nerve sheath tumour, suggesting it to be a neurofibroma (Figure 2).

Treatment

Routine pre-operative investigations were done. After obtaining Informed consent and getting pre-anesthetic evaluation done. The patient underwent excision biopsy of

the swelling, and the specimen was sent for histopathological examination (Figure 3).

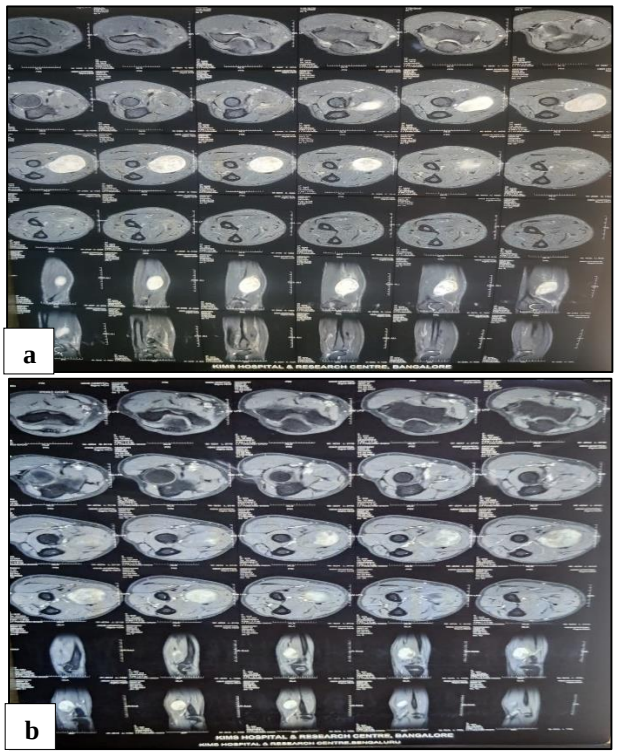

Figure 2: T2 weighted hyper intense lesion with target sign.

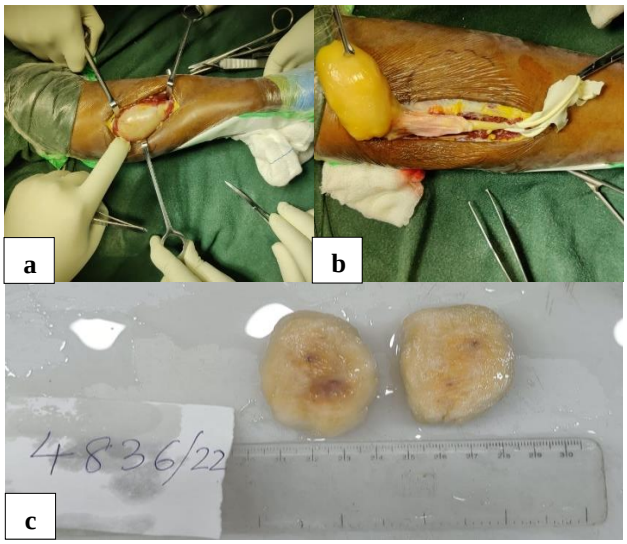

Figure 3: Intraoperative image with cut section showing hemmoraghic area.

Histopathology

Histopathological analysis confirmed the diagnosis of schwannoma, characterized by the presence of compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas, spindle-shaped cells arranged in a characteristic palisading pattern. No signs of malignancy were noted (Figure 4).

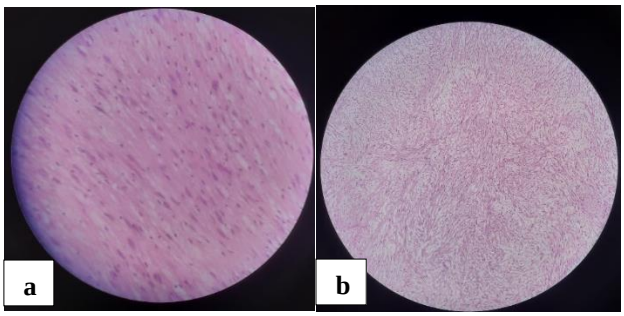

Figure 4: Characteristic features of Schwannomma – (a) Antoni A and (b) Antoni B areas.

DISCUSSION

Schwannomas are the most common benign tumors of peripheral nerves, often asymptomatic but can lead to significant morbidity due to mass effects with increase in size. It can result in spontaneous pain or pain after working, with paraesthesia and motor weakness as the main complaints.[6] The diagnosis may be challenging due to overlapping features with other soft tissue tumors. Although most of these tumors show a positive Tinel sign, paresthesia, and transverse mobility, their clinical identification is somehow subjective and thus are frequently misdiagnosed due to similarities with other soft tissue tumors as lipoma, fibroma, ganglion or xanthoma. Schwannomas are more likely to contain cysts, hemorrhage, fibrosis or calcification.[3]

MRI is the best imaging modality for diagnosing nerve sheath tumors, however, sometimes the differential diagnosis from other soft tissue tumors is difficult if they do not have specific signs. MRI appearances of schwannomas and neurofibromas are similar, and the detection of the nerve running into and out of a fusiform mass oriented along the line of the nerve is pathognomic for a nerve sheath tumor. MRI is the preferred imaging modality, displaying characteristic findings such as the "target sign" i.e. peripheral hyperintensity and central hypo-intensity on T2-weighted images due to outer myxoid, central fibrous tissue and continuity with a normal nerve. This aids in differentiating schwannomas from neurofibromas. Nerves tend to be displaced by the eccentric schwannomas, whereas the nerve is central to, or obliterated by, neurofibromas. The most reliable sign for schwannoma is its localization in continuity (eccentrically) with a normal nerve. MRI characteristic features show slightly increased signal intensity relative to muscle on T1WI, a thin peripheral rim of fat (split-fat sign) is very characteristic and may have subtle muscle atrophy distal to lesion.[7] On fluid-sensitive sequences schwannomas are hyperintense to muscle and may show a typical central low signal region (target sign) and multiple small ring-like structures (fascicular sign). Diffuse intense enhancement is typical.

In this case, the absence of regional lymphadenopathy, the characteristic clinical features, and histopathological confirmation underscored the diagnosis of schwannoma. Surgical excision is the mainstay of treatment and is usually curative, with low rates of recurrence and malignant transformation.

Despite being rare, tumors should be taken into account in the differential diagnosis of masses in the upper limbs. In this context, it is important to remember that Schwannoma, in these cases, is the most common tumor. The diagnosis and further treatment are not always prompt and accurate, which may lead to irreversible damage to the affected nerve with all the ensuing consequences. The simple removal of the tumour after careful dissection is generally enough since the recurrence and malignant transformation rates are low.

CONCLUSION

This case illustrates the importance of considering schwannomas in the differential diagnosis of forearm swellings, particularly when associated with neurological symptoms. This is a unique presentation of schwannoma distinguished by the presence of multiple hypopigmented patches and glove-and-stocking neuropathy, which are atypical for conventional schwannoma cases. Unlike most patients who present with isolated swelling or minimal neurological symptoms, this individual exhibited significant sensory disturbances and muscle wasting. The history of traumatic amputation further complicates the clinical picture, emphasizing the need for careful differential diagnosis. This case underscores the diverse manifestations of schwannomas and the importance of comprehensive evaluation in atypical presentations to ensure timely and appropriate management. Prompt diagnosis and surgical intervention can lead to favorable outcomes, preventing potential irreversible nerve damage.

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