

Multiple Schwannomas Of The Peripheral Nerve Or Schwannomatosis?

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INTRODUCTION:

Schwannomas are encapsulated, slow growing nerve sheath tumours which are almost always solitary. Only 3 to 4% are found to have multiple lesions¹. We present a case with 3 schwannomas fulfilling the criteria of schwannomatosis (two or more cutaneous or intradermal schwannomas with lack of evidence of vestibular tumor and no cutaneous manifestation)².

MATERIALS & METHODS:

A 76-year-old lady presented with multiple swellings over the right hand and forearm. These swellings started insidiously as pea sized and gradually enlarged over 5 years causing tingling like discomfort limiting her function. The three swellings were over the medial flexor forearm, ulnar border of wrist and palmar thenar measuring about 1cm x 2cm (Figure 1). They were firm, non tender, well defined, not attached to overlying skin or deeper muscle. Tinel's was positive over all the swellings. There were no palpable lymph nodes, no skin patches or skin freckling suggestive of neurofibromatosis.



Figure 1: Multiple swelling over forearm, wrist and thenar region.

RESULTS:

MRI showed four well defined lesions with intermediate signals on T1, high intensity on T2 and enhanced brightness on contrast (Figure 2). Two lesions arose from the median nerve and two from the ulnar nerve.

The swellings were surgically enucleated. The histopathological report showed well circumscribed masses of cellular areas of monomorphic spindle shaped cell with poorly defined eosinophilic cytoplasm and Antoni A and B cells consistent with schwannoma. One-month post operation, she experienced less discomfort of her right upper limb.



Figure 2 :1cm x 2cm hyperintense swelling medial to the distal ulna, homogenous and well encapsulated



Figure 3 : lobular , homogenous and well encapsulated Schwannomas

DISCUSSION:

A schwannoma almost always occurs as a solitary lesion except in neurofibromatosis. Schwannomatosis is now a distinct neurofibromatosis entity (NF 3)³ ie. benign, sporadic, non-hereditary characterized by anatomically localised multiple schwannomas without vestibular involvement.

CONCLUSION:

It is rare to see multiple nonvestibular schwannomas involving two peripheral nerves (median and ulnar) in a single limb, which we diagnosed as schwannomatosis.

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