



Segmental Schwannomatosis of Brachial Plexus- A Rare Case Report

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ABSTRACT

This paper reports the first case of multiple schwannomas from the trunks and divisions of brachial plexus on the same side in a patient without any other stigmata of neurofibromatosis i.e a case of segmental schwannomatosis from trunks and division of brachial plexus.

Our patient presented with swelling and pain in the left axilla of four months duration associated with tingling sensation in left upper limb on tapping the swelling. Magnetic Resonance Imaging of the left axilla revealed two well defined closely spaced lesions and suggested possibly schwannoma. Nerve Conduction Study of left upper limb showed left brachial plexopathy. Excision of the swellings were done via axillary and infraclavicular incisions with explained risk of nerve damage. Post operatively patient developed left wrist drop which improved with physiotherapy. The histopathology report came as schwannoma with degenerative changes.

KEYWORDS : Schwannoma, schwannomatosis, brachial plexus

Case presentation

32 year old house wife presented with painful swelling in the left axilla of four months duration. It was 6 x 5 cms firm oval tender swelling with smooth surface and no local rise of temperature. There was tingling sensation along medial border of left upper limb on tapping the swelling. She was otherwise in good health and had no significant past history or family history.

Upon investigation- MRI of the left axilla revealed two well defined closely spaced lesions along the trunks and divisions of left brachial plexus and suggested possibly schwannoma[figure 1]. Nerve conduction study showed left brachial plexopathy. USG both breasts suggested left axillary lymph nodes and FNAC from them showed only stromal fragments. MRI brain was normal.



FIGURE 1 Contrast MRI showing the schwannomas

Excision of the swellings were done via axillary and infraclavicular incisions with explained risk of nerve damage. There were two well encapsulated 5 x 5 x 4 and 5 x 5 x 3 cms swellings[figure 2]. Post operatively patient developed wrist drop which improved with phys-

iotherapy. The histopathology report came as schwannoma with degenerative changes.

Discussion

Schwannoma is the most common type of benign peripheral nerve sheath tumor which arises from the Schwann cell. [1]

They are slow growing and presents with swelling and pain or paresthesia upon percussing them. Most of them could be diagnosed clinically. The main differential diagnosis is neurofibroma. [2]

Schwannomatosis is a recently recognized form of neurofibromatosis characterized by multiple nonvestibular schwannomas without any stigmata of neurofibromatosis. Schwannomatosis is the third major form of neurofibromatosis and is a clinically and genetically distinct form.

The annual incidence is 0.58 cases per 1,000,000 persons.[3]

2.4 to 5% of patients requiring schwannoma excision have schwannomatosis. Approximately one third of patients with schwannomatosis have tumors limited to a single limb or segment of spine which is called as segmental schwannomatosis. [4]

Michael *et al* has described that a patient with definite schwannomatosis should have either age above 30 years and two or more non-intradural schwannomas, at least one with histological confirmation or one pathologically confirmed schwannoma plus a first-degree relative who meets the above criteria. [5]

Histopathological features of schwannoma are Verucay bodies and Antoni A and B areas, and immunohistochemical study is positive for S-100 protein.

MRI plays a critical role in diagnosing schwannomatosis. [6]

Treatment is surgical excision. Asymptomatic tumours can be monitored conservatively with serial MRI studies, usually at yearly interval. Long-term follow-up is essential as fresh lesions can occur any time. [7]



FIGURE 2 The swellings after excision

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