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SCHWANNOMA.

Orthopaedics

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ABSTRACT

Schwannomas are rare tumors. Since these tumors are derived from nerve sheaths and not from axons there is controversy regarding histogenesis and terminology. There is histological and histo-chemical evidence to consider that cells involved in the development of these tumors are schwannian cells and should be designated as schwannomas or neurilemomas, either benign or malignant. They are asymptomatic unless the lesion is large. The exact cause is not known.

KEYWORDS

Schwannoma, Neurilemomas, Neuromas, Neurolemomas.

INTRODUCTION:

Schwannoma is slow growing neurogenic tumor. Schwannoma referred as neurilemomas, neuromas, neurolemomas.[1] A schwannoma is a tumour which arises from schwann cells that grows in the sheaths of nerves in your peripheral nervous system. It is a slow growing neurogenic tumor.[2]It is usually benign and it could turn cancerous/malignant in a rare cases. Malignant schwannoma is so called soft tissue sarcomas. It usually affects 20-50 years of age and both the genders get affected equally. It can almost occur anywhere but flexor extremities are the commonest site.[3] Inter-muscular schwannoma arises from small nerve branch within the muscles.

Pathogenesis :

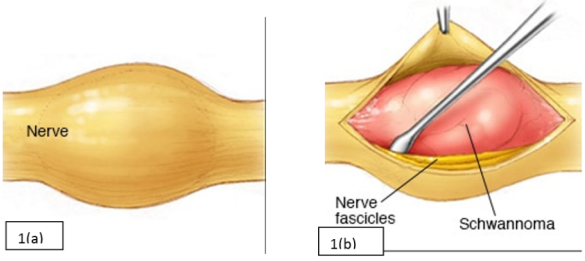
Schwannoma can occur spontaneously or from familial tumour syndrome like neurofibromatosis type 2 or schwannomatosis. It is a grade 1 tumour according to WHO.[4] The pathogenesis is loss of merlin, either by direct hereditary change involving the NF2 gene or inactivation of merlin.

Morphology :

There were no specific gross findings in a malignant schwannoma. Usually fusiform or oval tumours seen along the course of a nerve trunk. In cases of large proliferative lesion, it is sometimes enclosed by epineurium which is grey-white in color, well circumscribed and sometimes encapsulated.[5] The cut surface showed whitish, whorled or homogenous grey appearance with areas of hemorrhage and necrosis. A spindle shaped cell growth pattern with the cells arranged in tight wavy microscopic pattern.

Risk factors :

The exact cause is not known. Exposure of radiation is also a possible cause for schwannoma. Pre-existing neurofibromas, and neurofibromatosis type 1 (von Recklinghausen disease) is also the important risk factor.[6]



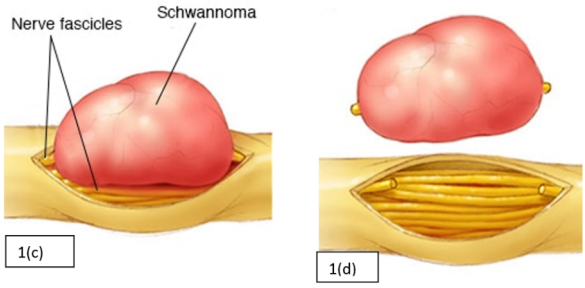


Fig 1 (a to d) : Steps for removal of schwannoma while taking care to preserve nerve fascicles that aren't affected by the tumor.

Symptoms :

They are asymptomatic unless the lesion is large. It may present as occasional pain in the area that is controlled by the affected nerve. Some other common symptoms include visible lump under the skin, a sharp aching or burning pain, a pins and needle sensation, muscle weakness, numbness.[8] Depending on the site the symptoms occur. Based on the severity, pain and neurological symptoms occur.

Investigations :

Diagnostic investigations include ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI) and fine needle aspiration (FNAC) .[9] MRI shows a well-encapsulated, round lesion within the inter-muscular space. The lesion shows homogeneous isointense signal intensity on T1WI, and heterogeneous hyperintense signal intensity on T2WI.

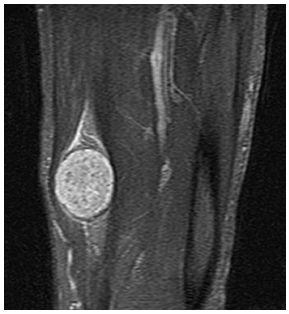


Fig 2 : MRI showing a fusiform soft tissue mass associated with the neuro- vascular bundle.

Management :

Intra capsular enucleation and complete tumor resection is the management for schwannomas.[10]

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