Schwannoma

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A Schwannoma is a tumor of the nervous system that grows from specialized cells called “Schwann cells”. It develops in the peripheral nervous system or nerve roots. These protective Swann cells are known to have a protective effect on the nerve cells of the nervous system. The schwannoma is often a benign tumor but in rare circumstances, they can become aggressive and present with a tumoral characteristic especially when they are involving the Sciatic nerve or the brachial and sacral plexus. The most common type is a vestibular schwannoma.

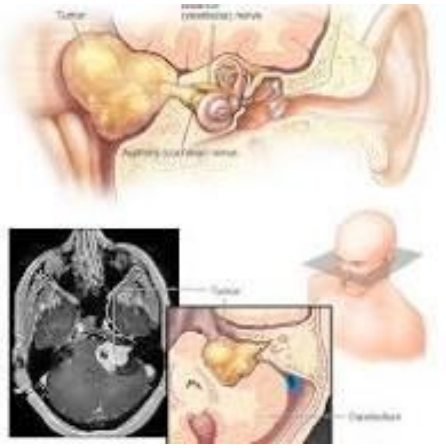
Although rare, a schwannoma is a pathology which affects less than 200,000 people. This is the most common of peripheral nerve tumor in the adult, but may occur at all age-group. They are almost always benign and non-cancerous. They are slow-growing and symptoms may differ depending on their size or their location but above all, the area where it is discovered in the body may dictate their influence. They are generally slow growing.

The Schwann cells assist in this peripheral nervous system. They are the glial cells that support neurons in the peripheral nervous system (PNS) and produce the myelin sheath that insulates axons, including nerves that travel from the spinal cord to the brain while allowing the signal to be carried to the rest of the body.

An area of predilection for a schwannoma is at the junction of the brain and the inner ear (vestibular or acoustic schwannoma) in 60% of cases, while one may encounter the malignant or cancerous form of Schwannoma occasionally around the sciatic nerve and occasionally the brachial or the sacral plexus. Another name generally given to these tumors are “Neurilemmomas or Neuromas”. In face of a malignant schwannoma, the litterature refer to a soft tissue sarcoma. It is generally accepted that schwannomas are solitary in 90% of cases.

Let us review the different types of schwannomas:

1- A Vestibular (acoustic) schwannoma affects generally people between the ages of 50 and 60 years, but rarely seen in children. They are seen randomly in healthy people but may also be associated with genetic conditions like Neurofibromatosis 2(NF2), schannomatosis or Carney complex. The schwannomas can be multiple in such pathology. They grow slowly and may exist for year before becoming symptomatic. They grow slowly and symptoms may vary. A lump can be visible and become tender at pressure, with muscle weakness or occasionally present with numbness but in the inner ear, the acoustic schwannoma affects the nerve of the inner ear causing hearing loss I the affected ear, ringing ear (tinnitus) and dizziness.



Acoustic or vestibular schwannoma

A schwannoma may be considered as a brain tumor or discovered elsewhere in the body itself. The most common type is in fact the vestibular or acoustic form of schwannoma. People may experience burning sensation, aching, ringing (tinnitus) in the ear. If the facial nerve is involved, facial paralysis can be seen as well as problems with eye motion, taste sensation and eye motion depending on the site of involvement on the trigeminal nerve.

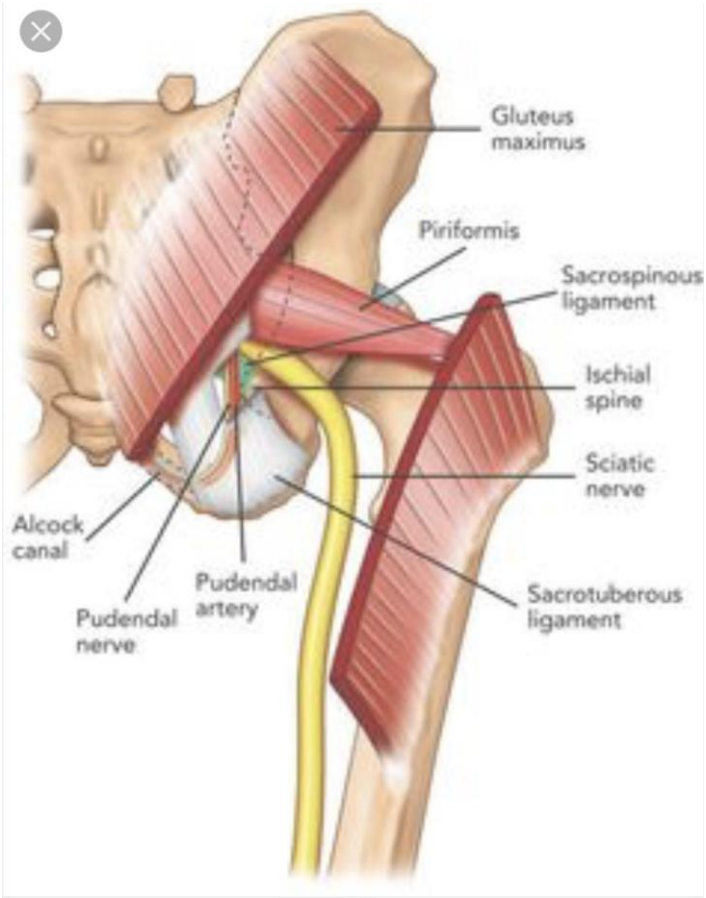
Dizziness, hearing loss, facial paralysis with difficulty in eye motion as well as in swallowing can represent some striking side effects for any lesion localized in the face. Inversely, lesions will involve the peripheral nerves especially at the level of the brachial or the sacral plexus, but the most disabling site I have observed in my practice, deals with the sciatic nerve itself. A case of

schwannoma involving this nerve at the level of the sciatic notch will be presented.

2-Symptoms related to schwannomas along the sciatic nerve can mimic disc herniation and low back pain with radicular symptoms to the lower extremity. (90% of those cases occur sporadically although they may be also associated to the Carney complex, neurofibromatosis 2(NF2) or schannomatosis. Studies have shown that the NF2 gene on the chromosome 22 plays an essential role in the development of a schwannoma. Fortunately, only 10% of the cases may have such genetic association.

A schwannoma can be present for many years, growing slowly and silently without becoming apparent.

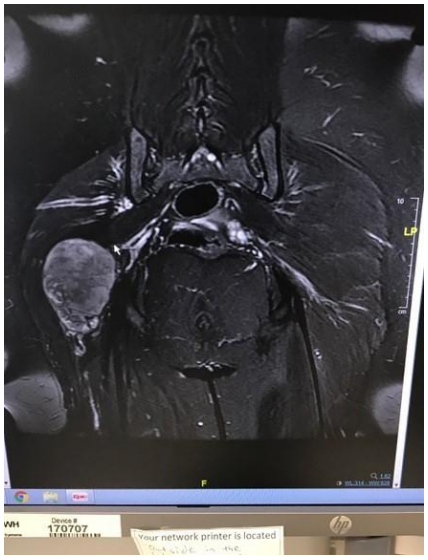
Sciatic nerve at the sciatic notch



Once a lesion becomes symptomatic, it may need to be investigated with imaging studies. Rarely, a schwannoma can be discovered incidentally, while a study is performed for another reason. Studies like Ultrasound, Computed tomography (CT), Magnetic resonance imaging (MRI) are as many tools available in the armamentarium of any examining physician to investigate such lesion.

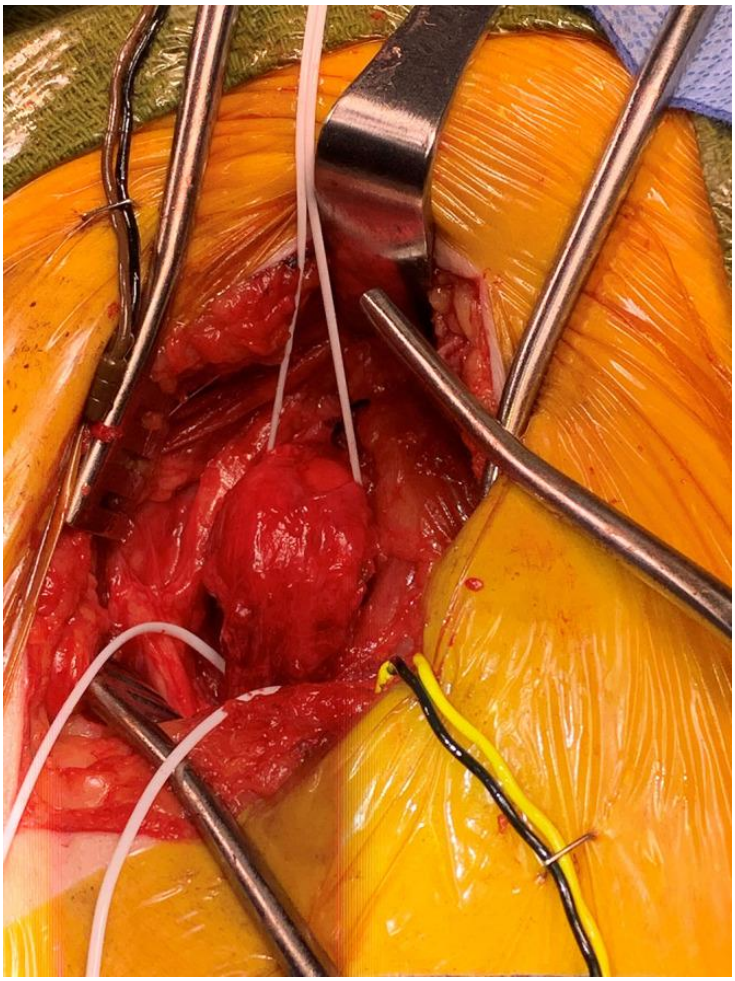
An interesting case is presented in a 40-year-old, young lady who started experiencing low back pain occasionally radiation to the right buttock twenty years ago. Often she will find uncomfortable to sit or lay down for a longtime in a certain position, without experiencing tingling sensation or numbness to the right lower extremity. She became pregnant in her twenties and the symptoms gradually increased in intensity bringing numbness and radicular symptoms to the right lower extremity as well as right buttock pain. Her first pregnancy has seen an increase in the symptoms with additional giving way episodes to the right lower extremity.

Investigations were carried out with MRI studies and the presence of a large lesion of the right sciatic nerve at the level of the sciatic notch was discovered. An extensive approach and dissection to remove the lesion were planned by two surgical teams an orthopedic team and a neuro-surgical team. It has permitted a complete excision of a large lesion of the sciatic nerve at the notch. Pathological diagnosis revealed the existence of a large “Sciatic nerve schwannoma”.



Large schwannoma at the sciatic notch

By now, she is G2P2 and has delivered 2 healthy children aged respectively 17 and 15. She has no more buttock pain but has experienced residual radicular symptoms in the peroneal nerve distribution which has imposed an ablation procedure of the right peroneal nerve at the tibio-peroneal bifurcation around the right fibular head. She has no more buttock pain and radicular symptoms to the right lower extremity described as phantom leg syndrome and has been able to wear ¾ inches shoed.

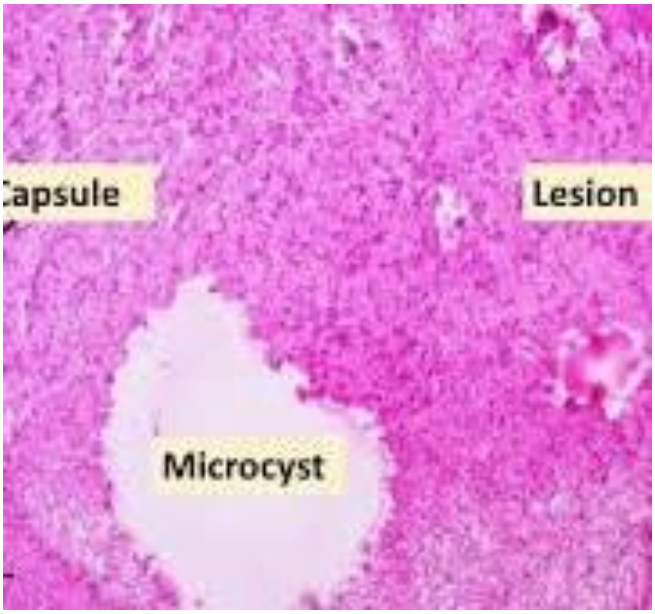


Operative view of the sciatic notch exposing a large Schwannoma.

In the right lower extremity of our patient above presented, one can understand the reason for such radicular symptoms just by observing the size and the position of such lesion in the sciatic notch. The lesion even appears to

be intra-articular requesting the expertise of the two expert-teams of specialists to resolve the problem. A schwannoma may involve other peripheral nerves in the body, producing pain out of proportion as well as an inability to ambulate or move the extremity or even sit down.

In brief, dealing with any schwannoma, we must consider the location and the size of the lesion. In the acoustic form, symptoms of hearing loss, dizziness and, imbalance or the presence of facial paralysis and pain, trouble in swallowing, numbness and tingling of the face, as well as trouble with eyes motion with facial paralysis, trouble in swallowing, tingling and numbness of the face may be striking.



Microscopy of a Schwannomas lesion

In any suspicion of a schwannoma, CT Scans and MRI may be helpful to define the lesion size and its location. Rarely, such tumor may spread to other parts of the body, forming “metastases” and rendering the lesion to become “malignant”. A biopsy will be always necessary to asset the diagnosis.

Although, a schwannoma is generally considered as a slow growing tumor requiring a wait and watch attitude, we should expect that once such lesion is discovered and a diagnosis made, to be able to offer treatment to the patient through an exploration and a surgical excision. Rarely, local radiation has

been offered to eliminate any chance of recurrences especially after the discovery of a high number of large tumoral or deep lesions, or metastases. Others may also consider any tumor originating also from one of the upper extremity long nerves. Such criteria are accepted by experts in the field to suggest additional "post-operative radiation therapy" in the goal of preventing any possible recurrences. "Stereotactic radiosurgery" (SRS) uses many precisely focused radiation beams to destroy any residual tumoral cells.

Follow-up is strongly suggested in such cases but although, a special attention should be given to patients with Neurofibromatosis type 2 (NF2) which are strongly associated to schannomatosis and Carney Complex in 10% of the cases. Finally, the presence of any malignant (cancerous) schwannomas may be also approached with Immunotherapy and possibly chemotherapy.

The prognosis is good following treatment of schwannomas, and the majority of such cases do not recur. I would like to take the opportunity to dedicate this paper to "CJ" for her suffering in dealing with such a large and rare lesion involving her right kip joint (sciatic notch), hoping that the definitive treatment offered will keep her free of any local or distant recurrences. So far, more than twenty five years after surgical treatment, she remains free of disease. Unfortunately, recurrences can't be prevented, especially when they happen randomly, for unknown reasons.

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References:

1- **MacCollin M, Chiocca EA, Evans DG, et al.: Diagnostic criteria for schwannomatosis. Neurology 64: 1838-1845. 2005.**
2- **Ogose A, Hotta T, Morita T et al.: Tumor of peripheral nerves: correlation of symptoms, clinical signs, imagins and histologic diagnosis. Skeltal Radiol 28: 1830188. 1999.**
3- **Das Gupta T, Brasfield RD: Solitary malignant schwannoma (Neurilemmoma). Cancer 24:213-235. 1964.**

4- **Skovronsky DM, Oberholtzer JC Pathologic classification of peripheral nerve tumors. Neurolog Clin N Am 15:213-257. 2004.**
5- **Varma DGK, Moulopoulos A, Sara AS, et al: MR findings in primary retroperitoneal nerve sheath tumors. J Comput Assist Tomogr 16:448-453.**
6- **Kim DH. Murovic JA, Tiel RL, et al: Operative outcomes of 546 Louisiana State University Health Sciences Center peripheral nerve tumors. Neurosurg Clin N Am 15:971-977, 2004.**
7- **Oontz NA, Wiens AL, Agarwal A, Hingtgen CM, Emerson RE, Morsier KM. : Schwannomatosis: the overlooked neurofibromatosis? AM J Roentgenol 2013 Jun. pp. 646-653.**
8- **Blanchard C, Dam-Hieu P, Zagnoli F, Bellard S: Chronic sciatic pain caused by sciatic nerve schwannoma. Rev Med Interne. 2008; 29:748-750.**
9- **Ghaly RF. A posterior tibial nerve Neurilemmoma unrecognized for 10 years: case report: Neurosurgery. 2001; 48:668-672.**