Schwannoma and Schwannomatosis

Schwannomas are benign nerve sheath tumors. They arise from the supportive tissue within the nerve itself. As these tumors grow, they displaced and compress adjacent nerve fascicles within the nerve. This causes pain, weakness, and numbness. Schwannomas usually are solitary and occur sporadically; however, in a some patients they can be part of a hereditary disorder (e.g., neurofibromatosis or schwannomatosis).

Diagnosis

Because these tumors can occur in any nerve in the body, symptoms are variable. In general, schwannomas can be asymptomatic, cause progressive pain, weakness, tingling, and numbness, or present as a mass in the neck or extremities. Tapping on the mass often causes electrical pain to shoot down the extremity (i.e., Hoffman-Tinel's sign). These tumors are classically mobile perpendicular to the parent nerve, but not along the trajectory of the nerve. Although one can often suspect the diagnosis of a schwannoma on history and examination, it is confirmed with MRI, with and without contrast. It is usually not possible to differentiate a schwannoma from a solitary neurofibroma using MRI preoperatively. High-resolution ultrasound may also be used to evaluate a schwannoma. Definitive diagnosis is obtained after these tumors are removed, or during tumor resection. In general, needle biopsy is not recommended. Preoperative electrodiagnostic testing is often not required.

Treatment

Small, asymptomatic schwannomas, especially in older patients, may be observed with serial MRIs. If weakness, numbness, or pain is present, then microsurgery to remove these lesions is recommended. Although radiosurgery may be an option for schwannomas in the head or spine, for the extremities this is currently not a viable option. Pre- or post-operative neuropathic pain caused by a schwannoma may be treated with oral medication (e.g., gabapentin). Surgery is the mainstay of schwannoma treatment. The nerve is exposed under general anesthesia. Using a microscope and intraoperative electrophysiological monitoring, the nerve is opened and tumor is carefully removed from the nearby functional nerve fascicles. The tumor usually arises from one small nerve fascicle that has been made non-functional by the tumor-this fascicle is removed with the tumor. The remaining nerve fascicles are preserved, thereby maintaining the nerve's function. Surgery is efficacious in resolving pain, weakness, and numbness in the majority (80-90%) of patients. Post-operative physical therapy is recommended is there is motor weakness or painful numbness.

Complications

Besides the normal risks of surgery, including reactions to anesthesia and infection, patients who undergo schwannoma removal are at risk for paralysis, numbness, and worsened pain. Although these complications rarely occur, they are possible. These risks depend on multiple factors, including size and location of the tumor, as well as preoperative clinical status of the patient.

Outcome

With complete tumor removal, the chance of recurrence is minimal, perhaps a few percent over the patient's lifetime. Therefore, serial postoperative imaging is not required. Resolution of preoperative pain and/or weakness occurs in about 85% of patients, however, it may take weeks to months to occur, depending on how long it was present before surgery. The risk of motor weakness from the resection is low, however the chance of a permanent or temporary sensory deficit is relatively high (15-20%). Neuropathic pain is perhaps the most concerning complication, which can occur both from surgery, or in patients with chronic pain secondary to an untreated tumor. The chance of problematic neuropathic pain after surgery is estimated to be about 5%.