

Malignant Schwannoma

The malignant schwannoma is a high-grade spindle cell sarcoma arising from the nerve sheathes of peripheral nerves. It can arise denovo from a normal-appearing peripheral nerve but is more likely to arise from a solitary neurofibroma. Malignant schwannoma from a peripheral solitary neurofibroma occurs more often in people over the age of 40 years. Those patients have a 75 per cent chance of survival at five years. Malignant schwannoma from neurofibromatosis (a 10 per cent incidence) occurs more often in a young age group and the chance for survival at five years is reduced to 30 per cent. The malignant schwannoma is usually larger than 5 centimeters and occurs typically in spinal roots or in the larger proximal nerves such as the sciatic nerve. It presents as a painful mass in patients with neurofibromatosis and there may be neurological deficits distal to the involved area. There may also be a café-au-lait spot in the overlying skin. The lesions are best picked up by magnetic resonance imaging. The tumor is usually treated by a wide resection, including the entire nerve from which it originates, following which local radiation therapy is recommended to reduce the chance of local recurrence. Adjuvant chemotherapy is advised even though its benefit is controversial.