Neurothekeomas are benign connective tissue tumors probably of nerve sheath origin. Making diagnosis is often difficult, because of many histological similar looking tumors. Immunostaining of S-100 protein is a helpful method for differentiation. We report a case of subungual neurothekeoma affecting the little toe, which is to our knowledge the first to be described in the literature. In spite of an incomplete excision of the tumor with tails reaching to the base of the specimen, no recurrence after 1-year follow-up was observed.