Case Study

Ancient Schwannoma with Cyst Formation and Degenerative Changes in Sciatic Nerve: A Case Report

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ABSTRACT

Schwannomas of the sciatic nerve are rare and well-delineated tumors, excentrically located on the nerve. Ancient schwannomas can vary from firm, solid masses to fluctuant cysts. Here we present a case of 67 year old woman with swelling in thigh. Clinical diagnosis was misleading and was diagnosed as intermuscular lipoma. Only with histology, the actual report of ancient schwannoma with cyst formation and degenerative changes of sciatic nerve was given.

Key words: ancient schwannoma, sciatic nerve.

1. INTRODUCTION

Schwannoma, also called neurinoma or neurilemmoma, is a benign peripheral nerve sheath tumor. They generally appear as solitary lesions. Ancient schwannomas can vary from firm, solid masses to fluctuant cysts. Schwannomas of the sciatic nerve are rare and well-delineated tumors, excentrically located on the nerve. Here, a case of an ancient schwannoma of sciatic nerve is presented and literature on sciatic nerve schwannomas is reviewed.

2. CASE REPORT

A 67 year old woman presented with a slow growing, painful swelling in left thigh since one month. On physical examination 6x8 cm tender, soft, movable...
mass in the posterior aspect of thigh was found. No motor deficit was detected on neurological examination. A clinical diagnosis of intermuscular lipoma was made. The patient was from poor socioeconomic background, hence she did not agree for ultrasonography and MRI scan. FNAC was done. Cytology showed scattered mixed inflammatory cells, lymphocytes, macrophages, hemosiderin laden macrophages and neutrophils. No atypical cells were seen. The clinical laboratory findings were within normal limits. The patient agreed for excision of the lesion. Intraoperatively, the mass was found below muscle extenders of thigh, adherent with sciatic nerve, compressing it posteriorly. The surgical specimen was sent to pathology department for histological examination. Gross findings showed a well circumscribed cystic mass measuring 5x5x3cm. The external surface showed foci of hemorrhagic areas. Cut surface showed multiloculated cystic areas filled with blood and brown soft tissue. Foci of solid areas were also seen (figure 1). Microscopy showed, an encapsulated neoplasm with extensive cystic degeneration and interstitial hyalinization (figure 2). The tumor cells showed alternating cellular Antoni A and hypocellular Antoni B areas. Antoni A areas with tumor cells in short fascicles and focal nuclear palisading were seen (figure 3, 4). Antoni B areas with loosely textured hyalinised stroma were seen (figure 5). Ectatic irregularly shaped vessels, thrombosed blood vessels with surrounding hyalinization (figure 6), cystic spaces resembling dilated lymphatics were also seen. The tumor showed infiltration by large number of siderophages and histiocytes (figure 7). A few schwann cells showed nuclear atypia with large hyperchromatic nuclei, but lacked mitotic figures (figure 8). The tumor showed positive immunostaining for S 100. According to these histological findings, the diagnosis of an ancient schwannoma with cyst formation and degenerative changes was made.

3. DISCUSSION

Schwannomas are derived from Schwann cells of neuroectoderm. They serve for the formation of myelin sheaths of nerves that insulate nerve and facilitate the transmission of an impulse. Schwannomas most commonly occur in adults between 20 and 50 years of age, without distinction of gender, with an approximate one sex ratio. They generally appear as solitary lesions. Malignant transformation in schwannomas is rare, the risk of malignization being approximated at 18% in neurofibromatosis type 1, and 5% in schwannomas. Patients with von Recklinghausen disease carry a worse outcome.  

![Fig 1](image1.jpg)

Fig 1: Gross findings showed a cystic tumor filled with brownish fluid and foci of myxoid areas.

![Fig 2](image2.jpg)

Fig 2: Microscopy, Ectatic irregularly shaped vessels, thrombosed blood vessels with surrounding hyalinization, cystic spaces resembling dilated lymphatics were seen.

![Fig 3, 4](image3.jpg)

Fig 3, 4: Antoni A areas with Verocay bodies.
Histopathologically, five schwannoma variants have been described: common, plexiform, cellular, epithelioid, and ancient schwannomas. Schwannoma consists of two components, Antoni A areas are more organised and are hypercellular, and are composed of spindle cells arranged in short bundles or interlacing fascicles. Antoni B regions are hypocellular, less organised and contain more myxoid, loosely arranged tissue, with a high water content.

Ancient schwannomas are rare, encapsulated tumours of long duration and are benign in nature. The tumour is solitary and may grow to a large size before detection of notable degenerative changes. The term “ancient schwannoma” is used to describe a tumour that has undergone such changes, typified by relative loss of Antoni type A tissue, perivascular hyalinisation, calcification, cystic necrosis, haemorrhage and the presence of degenerative nuclei that may be misinterpreted as sarcomatous pleomorphism. The term “ancient neurilemmoma” was first suggested by Ackerman and Taylor in a review of 48 neurogenic tumours of the thorax. The cited authors reported 10 patients with tumours showing features similar to those of typical neurilemmomas, but differing in that significant tumour portions contained only a few cells within hyalinised matrices. They found that these features occurred in schwannomas of long duration, and hence coined the term “ancient schwannoma”.

Such tumours are characterised by diffuse hypocellular areas, relative loss of Antoni type A tissue, focal accumulations of hyaline material, calcification, cystic necrosis, haemorrhage and fatty degeneration. In addition, as the tumours are usually infiltrated by large numbers of siderophages and histiocytes, and display cellular degenerative changes, including nuclear atypia and pleomorphism, along with a tendency to nuclear palisading, malignancy may be erroneously diagnosed. On histological tumour examination, intense immunostaining for S100 protein suggests a neural origin and is helpful in diagnosis, especially of a totally cystic degenerated mass.

Schwannomas have a long subclinical course and their clinical presentation will be usually misleading. The most common clinical presentation of sciatic nerve schwannoma consists of a painful palpable mass. Other clinical symptoms include radicular and distal pains often distant from the lesion site, paraesthesia, hypoesthesia and rarely motor deficiencies. Hence in our case too, it was misdiagnosed as intermuscular lipoma clinically. Surgical excision is the treatment of choice. Schwannomas are theoretically removable since they repulse fascicular groups without
penetrating them thus allowing their enucleation while preserving nerve continuity.\(^9\)

The posterior tibial nerve at the tarsal sinus is the most frequently involved nerve of the lower limb. Involvement of the sciatic nerve is rare and represents less than one over 100 cases. The nerve might be affected by the tumor all along its course.\(^9\)

Unresponsive pain and an irrelevant spinal MRI exam must conduct investigations in searching other nerve pathology. EMG and ultrasound may, also be useful in establishing the diagnostic of sciatic schwannoma. Y Soo lee\(^1\) and collegues reported two cases of ancient schwannoma of thigh with MRI features, suggesting that the mass should have a fibrous capsule and split fat sign.

4. CONCLUSION

In conclusion, we present a case of an ancient schwannoma of sciatic nerve, the clinical diagnosis could be misleading and hence excision and histopathological examination is mandatory.

5. REFERENCES


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