Case Report

Ancient Schwannoma of Radial Nerve: A Rare Case Report

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Abstract

Introduction

Schwannoma is a benign tumor of nerve sheath origin. Ancient schwannomas were first described by Ackerman and Taylor in 1951. In the upper limb, the ulnar and median nerves are the primary nerves of flexor surfaces where schwannomas typically arise. The radial nerve is involved in just 7% of peripheral schwannomas.

Case Report

A 25-year-old male presented to the surgery OPD with a history of forearm swelling for 5 years. On ultrasonography (USG), there was a hypoechoic lesion in the anterior aspect of the forearm. Based on the long duration, clinical examination, and imaging, a diagnosis of benign nerve sheath tumor was made. Excision biopsy was performed, and a globular grey-white tissue piece measuring 2.5x1.5 cm was received in the Department of Pathology. Sections from the globular mass revealed a well-circumscribed tumor with oval to spindle cells randomly arranged within myxoid stroma. There were areas of marked degenerative changes with occasional interspersed hypocellular areas. Immunohistochemistry (IHC) was applied to further confirm the origin of these cells. These cells were positive for vimentin and S-100. The final diagnosis, based on histomorphological features and IHC, was given as ancient schwannoma.

Conclusion

Ancient schwannomas involving the radial nerve are very rare. They can be confused with malignant peripheral nerve sheath tumors on imaging due to degenerative features and nuclear pleomorphism. Histopathological examination along with IHC is crucial in the diagnosis of these rare tumors. We report the fourth case of an ancient schwannoma in the upper extremity involving the radial nerve.

Keywords:

Ancient schwannoma, neural tumor, radial nerve

Introduction

Schwannoma or neurilemmoma is a benign tumor of nerve sheath origin. This tumor can originate from any neuron that has a sheath of Schwann cells covering it, such as the autonomic, spinal, and cranial nerves (except the ocular and olfactory nerves) [1]. Schwannomas affecting the upper extremities comprise 5% of all soft tissue tumors [2]. They mostly affect the major flexor surface nerves such as the ulnar and median nerves. Only 7% of all peripheral schwannomas involve the radial nerve [3].

Although it may arise at any age, the peak incidence is usually seen between the third and sixth decades. There is no sex predilection [4]. Due to its low incidence, clinical signs and symptoms are sometimes mistaken for other soft tissue tumors such as ganglioma or tenosynovitis [5]. The majority of these tumors are seen as solitary masses with dimensions ranging from 1 to 2.5 cm. Their slow and non-infiltrative growth pattern makes them unique in their presentation, although pain and neurological problems are often seen when the tumor is larger than 2.5 cm [6, 7].

Ancient schwannoma is a rare variant of schwannoma. These tumors make up 0.8% of all soft tissue tumors. They are typically long-standing and show substantial degenerative alterations such as calcifications, cysts, hemorrhage, and nuclear atypia. These degenerative changes and pleomorphism can pose a diagnostic challenge due to their similarity with sarcoma [8]. Thus, for diagnosing ancient schwannoma, histological examination along with IHC remains the gold standard.

Case Report

A 25-year-old male presented to the surgery OPD with a history of forearm swelling for 5 years, which increased in size over the last six months. On physical examination, a 2.0 x 1.0 cm non-tender, firm, round, non-mobile mass was found in the anterior aspect of the forearm. The patient had no neurological deficit. On USG, there was a hypoechoic lesion in the anterolateral aspect of the forearm along the course of the radial nerve. Based on the long duration, clinical examination, and imaging, a diagnosis of benign nerve sheath tumour was made.

An excision biopsy was performed, and a globular grey-white tissue piece measuring 2.5 x 1.5 cm was received in the department of pathology for histopathological examination. On cut section, it was brownish-white in appearance.

Sections examined revealed a well-circumscribed tumor with oval to spindle cells randomly arranged within a myxoid stroma. Some of these cells had bilobed and multilobed nuclei, with individual nuclei showing moderate pleomorphism at places. There were areas of marked degenerative changes, including cyst formation, hyalinisation, and hemorrhage, with occasional interspersed hypocellular areas (Figures 1 and 2).

IHC was applied to further confirm the origin of these cells. These cells were positive for vimentin and S-100 and negative for cytokeratin, epithelial membrane antigen (EMA), HMB 45, and desmin (Figures 3 and 4). The final diagnosis, based on histomorphological features and IHC, was given as ancient schwannoma.

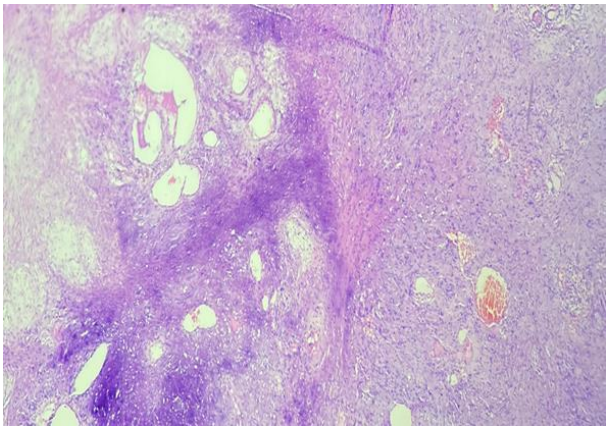

Figure 1: Microphotograph shows areas of cyst formation, myxoid change, and calcification (H&E, 40X).

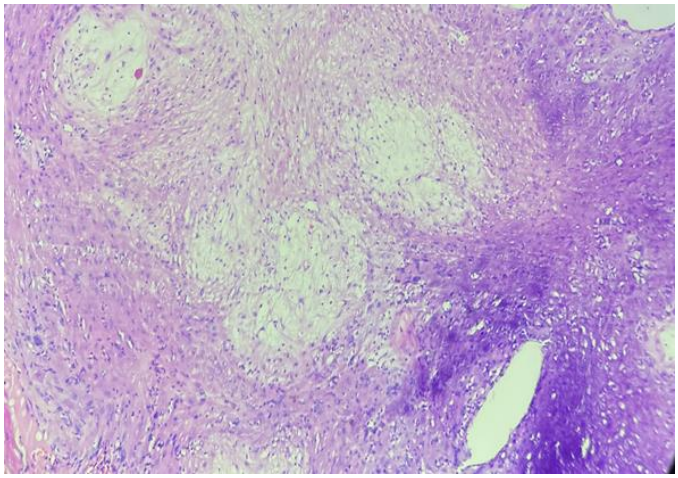

Figure 2: Microphotograph shows spindle cells exhibiting moderate pleomorphism with areas of cyst formation, myxoid change, and calcification (H&E, 100X).

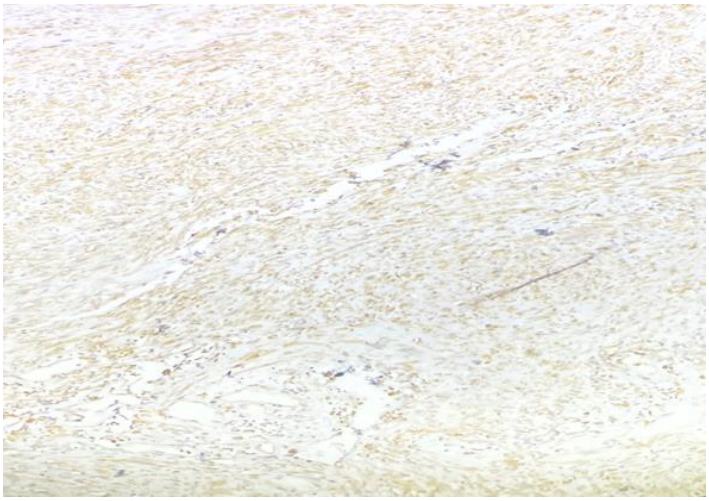

Figure 3: Micrograph showing spindle cells positive for Vimentin (Vimentin IHC, 100X).

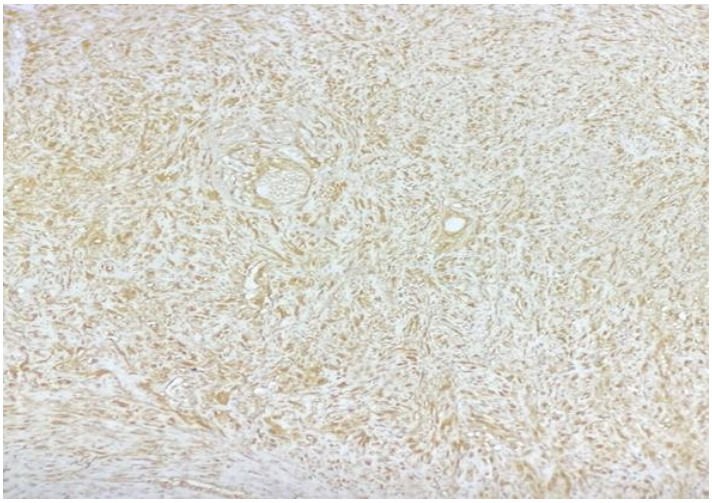

Figure 4: Micrograph showing spindle cells positive for S-100 (S-100 IHC, 100X).

Discussion

The incidence of Schwannoma in eastern countries is 5% in adults and 2% in children [9]. Ackerman and Taylor originally described ancient schwannomas in 1951 [10]. It is a solitary tumor arising from nerve sheaths, seen in individuals between 20 to 50 years of age. They can be asymptomatic or painful. Due to their slow development, there are no motor deficiencies as it enables the nerve to adjust to pressure effects [11]. It is characterized by cystic necrosis, stromal edema, xanthomatous change, fibrosis, perivascular hyalinization, calcification, and degenerative nuclei with pleomorphism and hyperchromasia. All these features can probe the diagnosis of malignancy.

In a study by Bhat et al., the first case was of a 62-year-old man with painful swellings on his left hand for six years. Despite paresthesia in the thumb, there was no motor or sensory loss. USG revealed hypoechoic lesions diagnosed as benign nerve sheath tumors. These were excised, revealing ancient schwannoma on histopathology. The patient remained symptom-free after one year. The second case was of a 56-year-old man with a history of painful swelling on his right arm for a year, causing paresthesia along the radial nerve distribution. Imaging and nerve conduction studies indicated a benign tumor along the radial nerve, which was excised with affected nerve fascicles, followed by repair. Pathological examination confirmed ancient schwannoma. At one-year follow-up, the patient experienced mild tingling but no neurological deficits [3].

In another study by Phan D et al., a 40-year-old man noticed a growing lump in his right forearm a year ago, which was initially small and painless. A biopsy confirmed it as a benign neural tumor. However, due to a significant increase in its size, surgical removal was advised. Histopathological analysis revealed an encapsulated tumor with ancient changes, including hemorrhage and strong S-100 positivity, consistent with ancient schwannoma [12].

Other single case reports have been published, like the study by Malizos et al. [11] and Shubhashraj et al. [13], who reported cases of ancient schwannoma of the medial nerve and mental nerve, respectively. In each of these cases, histopathological examination supplemented by IHC was pivotal in reaching the conclusive diagnosis of ancient schwannoma. This underscores the critical role of IHC in confirming the ultimate diagnosis.

In the present case, the patient had a history of swelling in the forearm, which recently showed an increase in size. Its microscopy revealed oval to spindle tumor cells randomly arranged in myxoid stroma and various degenerative changes, such as cyst formation, hyalinization, and hemorrhage, which can be misinterpreted as malignant tumors such as malignant fibrous histiocytoma (MFH), malignant peripheral nerve sheath tumor (MPNST), and synovial sarcoma. As there was a lack of characteristic histology of the fascicular growth pattern of malignant peripheral nerve sheath tumor and synovial sarcoma, both were ruled out. Further, various IHC stains were done, of which vimentin and S-100 were positive, helping to reach the final diagnosis in the present case. MFH is negative for S-100, thus supporting the neural origin of the tumor in the present case and probing the diagnosis of ancient schwannoma. A diagnosis of ancient schwannoma of the radial nerve, although rare, should always be kept as a differential diagnosis for forearm swellings to prevent overdiagnosis and overtreatment in a benign tumor.

Conclusion

Ancient schwannoma of the radial nerve is exceptionally rare. Thorough history taking, clinical and radiological assessments, histological analysis, and IHC are indispensable for identifying these uncommon tumors from their differential diagnosis, which can help prevent unnecessary sacrifice of the parent nerve. This case report offers valuable insights into this condition, shedding light on its rarity, potential predisposing factors, and differential diagnosis. Furthermore, distinguishing ancient schwannomas

from malignant tumors carries significant implications, as misidentification could lead to inappropriate treatment strategies, potentially compromising patient outcomes. Thus, meticulous diagnostic processes are essential to ensure proper management and prognosis. To the best of our knowledge, only three cases have been described in the literature; we hereby report the fourth case of an ancient schwannoma affecting the radial nerve in the upper extremities.

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Competing Interest: None Declared

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