Case Report

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ANCIENT SCHWANNOMA INVOLVING THE SUPERFICIAL PERONIAL NERVE: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

INTRODUCTION: Schwannomas are benign ,encapsulated,slow-Growing and usually solitary tumors originating from Schwann cells of the peripheral nerve sheath. Histological findings are these seen as in conventional schwannomas, but ancient schwannomas additionally demonstrate cystic, hemorrhagic changes and degenerative nuclei with pleomorphism and hyperchromasia. Due to the nuclear atypia, and cystic degeneration, ancient schwannomas might be confused with malignant tumors on histology and imaging, leading to a radical surgical approach .Schwannomas of the superficial peroneal nerves are very rare, We present a rare case of an ancient schwannoma involving the Superficial Peroneal nerve of right lower leg. The tumor slowly grew up within one year and become symptomatic with local pain, mild numbness in the distribution of the Peroneal nerve in the right leg. The tumor was successfully removed by separating it from the nerve fascicles to negative margins. Post-operatively local symptoms relieved but minor sensory loss in the Peroneal nerve distribution in the leg was noticed which improved in the following six months. Ancient schwannomas can be misdiagnosed as sarcomas due to specific imaging and histologic findings. Patients' physical examination, history and fine radiologic and pathology features should be cautiously interpreted in order to achieve correct diagnosis and avoid unnecessary wide tumor excisions.

PRESENTATION OF CASE: A 72-yr old male patient presented to us with complaints of swelling over the anterolateral aspect of right leg lower third region with pain and numbness since 1yr. Swelling was in initial size being that of pea nut which gradually progressed to about 6 x 3 x 2 cm at the time of presentation. Patient did not give any history of trauma/prick injury Diagnosis of a benign peripheral nerve tumor was achieved and the patient was treated by surgical excision of the lesion.

DISCUSSION: The intermittent symptomatology presentation on this case suggest a mechanical compression etiology, allied to classical pain and paresthesia often exhibited by this kind of the tumor. An intercompartmental pressure elevation could explain why the symptoms disclosed an episodic pattern, due to a constricted ,in closed nerve.

CONCLUSION: We describe a rare case of a patient with an unusual superficial peroneal nerve Schwannoma clinical presentation.Literature on this topic is scarce and,therefore,this case report intends to add further data about this kind of lesion.

KEYWORDS

Pleomorphism, Hyperchromasia, Creatinine Kinase, Aldolase Levels

ARTICLE HISTORY

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Introduction

Scwannomas are benign, encapsulated tumours arising from the Schwann cells of the nerve sheath, and were originally described by Stout in 1935 [1]. Batsakis more accurately described these tumours as neurilemmomas, referring to the cells of origin [2]. Approximately 25-40% of all neurilemmomas occur in the head and neck region [3]. The acoustic nerve is the most frequent site involved. Other locations described in the literature include the scalp, oral cavity, pharynx, larynx, parotid gland, middle ear and sinonasal tract. [4, 5]. Although they represent the commonest benign peripheral nerve sheath tumors,the occurrence on the lower limbs account for1% of all cases(.6) In the extremeties most commonly found in the deep tissues of the foot (7).Reports of Schwannomas arising specifically from the superficial peroneal nerve are exceptionally rare.(8)

Case report-

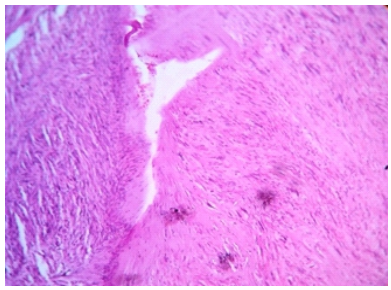
A 72-yr old male patient presented to us with complaints of swelling over the anterolateral aspect of right leg lower third region with pain and numbness since 1yr. Swelling was in initial size being that of pea nut which gradually progressed to about 6 x 3 x 2 cm at the time of presentation. Patient did not give any history of trauma/prick injury.. No history of constitutional symptoms and any other similar swellings in the body. On examination proper, swelling extended from the 5cm from tip of lateral malleolus to 8cm . Swelling was ovoid in shape with Small wound over anterior aspect of swelling with serous discharge. Swelling was firm in consistency with mild tenderness and no local rise of temperature. It was nonreducible and non-translucent, immobile in both horizontal and vertical direction. Sensations are diminished over dorsum of right foot. No motor weakness present. Movements at right ankle joint are normal.

X-ray revealed well defines lesion in the soft tissue with erosion of underlying bone..Serum calcium and serum phosphorus was 10.2 mg/dl and 3.0 mg/dl respectively. Serum ALP (127 IU/l), parathyroid hormone levels, creatinine kinase, aldolase levels, ANA, Vitamin D levels, 24 hours urinary calcium and inorganic phosphate were all within normal limits. Ultrasound of the swelling confirmed large well defined multiple lobulated, hypoechoeic lesion with few echogenic areas and fibrous septa in it. MRI of the lower third leg with ankle was done which revealed mulitlobulated, multiseptated soft tissue lesion in the lower aspect of anterolateral muscle compartment of right leg suggestive of schwannoma and hence excision biopsy was planned for confirmative diagnosis and definitive treatment.

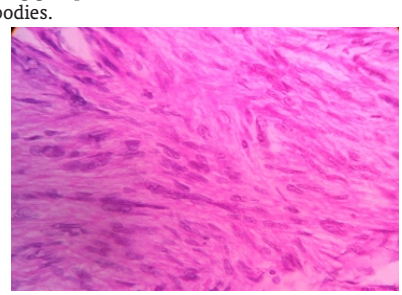
Gross examination of the biopsy specimen revealed grayish white mass measuring 5x3x1 cm, along with multiple tiny masss altogether measuring 2x1x1 cm. cut section shows homogenous grey white nodular areas.

M/E-composed of slender spindled cells in a loose, fibrous background with focal areas of hypercellularity alternating with hypocellular area. There were focal areas of pleomorphism of the cells ,hyperchromatic

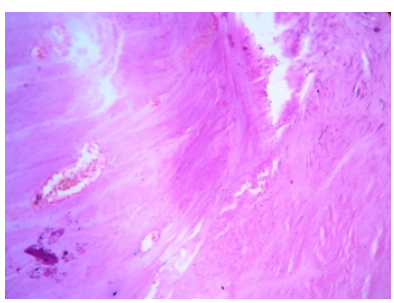
nuclei and chronic inflammation. There was no mitotic activity present. There was hayalinzed areas present.At some places myxomatous area was also present. The tumor cells were strongly S100 positive.A diagnosis of ancient schwannoma was made and the tumour was excised under general anaesthesia. Early follow up showed no signs of local recurrence of the tumour.



H and E staining of the specimen showing tumor cells composed of proliferating groups of Schwann cell nuclei in Antoni A tissue forming Verocay bodies.



H &E section showing nuclear pleomorphisim & few hyperchromatic nuclei



H&E section shows hyalinization and haemorrhge.

Discussion-The term ancient schwannoma was first introduced by Ackerman and Taylor in 1951 [9]. They described pathologic characteristics such as large areas of hyalinized matrix, increased hypercellularity with nuclear pleomorphism, and hyperchromatism. Ackerman and Taylor stated that these features occurred in schwannomas of long duration, leading to the term ancient schwannoma.

Schwannomas most often occur in the fourth and fifth decade of life and seem to have a1.6:1 female predilection.(10) They have been found to be present in such varied locations as the brachial plexus and the sciatic nerve.Rarely,Schwannomas can be found in the leg or in the foot and ankle region.(11). The presence of hypercellularity and atypia may lead to the misdiagnosis of these lesions as sarcomas. Dahl reported 6 cases out of 11 that have been misdiagnosed as sarcomas[12].The term ancient schwannoma, which is not an indicator of malignancy, is used to describe an old schwannoma that has undergone degenerative changes over time (this variant of schwannoma is also rare). Degenerative changes that characterize an ancient schwannoma include interstitial hyalinization, cyst formation, calcification and haemorrhage, along with degenerative nuclear atypia, but without any mitotic activity[13,14,15].

The evaluation of a patient with a suspected pathologic nerve condition broadly includes history,physical examination,and ancillary studies including several imaging modalities which provide valuable information in these disorders.Plain films may not reveal any changes whereas computedtomography(CT),magnetic resonance (MR),especially MRneurography,may display a peripheral nerve tumor in a more detailed manner.Additional diagnostic tests, including electromyography(EMG) and nerve conduction study (NCS),evaluate neuromuscular function to assess denervation, preservation of motor units,or conduction loss.(16)

On histological tumor examination, intense immunostaining for S100 protein suggests a neural origin and is helpful in diagnosis, especially of a totally cystic degenerated mass [17, 18]. Because the tumor contains cystic areas, ancient schwannoma has been radiologically misdiagnosed as other tumor types, such as malignant fibrous histiocytoma, malignant peripheral nerve sheath tumor,liposarcoma, synovial sarcoma or hemangiopericytoma. However, only a few reports have dealt with the radiological features of ancient schwannoma in the thigh because the tumor is so rarely encountered [19].

Conclusion-

These benign neural sheath neoplasms are infrequent in the lower extremities according to the literature. In the differential diagnosis of non traumatic leg pain, benign tumors, particularly Schwannomas of the peroneal nerves should be considered. Imaging studies, mainly magnetic resonance, are quite helpful in achieving a correct diagnose. Complete surgical resection by intralesional enucleation results in cure and recurrences are uncommon. We describe a rare case of a patient with an unusual superficial peroneal nerve Schwannoma clinical presentation.Literature on this topic is scarce and,therefore,this case report intends to add further data about this kind of lesion.This case evinces a non-reported clinical presentation, on an infrequent location, for this specific neoplastic disorder, and so depicts an added value to the knowledge of these cases in the literature.

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