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Ancient Schwannoma of the Distal Ulnar Nerve: A Rare Presentation

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Abstract

We describe the first rare case of a 3 year long standing ancient schwannoma in a 67 year old male involving the ulnar nerve just proximal to the Guyon's canal in the flexor aspect of the right distal forearm with a positive Tinel's sign and sensory deficit. He was managed with marginal resection and meticulous fascicle dissection and had an excellent clinical outcome with no neurovascular deficits at a 6 month follow-up. Histological findings, (both macro and microscopic) revealed a biphasic variegated pattern of hyper and hypo cellular regions with marked nuclear pleomorphism enclosed in a well encapsulated sheath.

Keywords: Ancient schwannoma; Ulnar nerve; Elderly; Extremity; Tinel's sign; Hyper and Hypocellular; Pleomorphism

Introduction

Ancient schwannomas are a rare entity in literature and were first described by Ackerman and Taylor in 1951 [1,2]. They are so named owing to the elderly age of the patients and long standing nature which eventually undergo degenerative changes [1-4]. There has been an increased incidence of early misdiagnosis of these tumors as sarcomas or soft tissue tumors given the age of presentation, location and microscopic degenerative changes with nuclear atypical [5,6]. Literature reports a more frequent presentation in the head and neck region, trunk, retro peritoneum, pelvis and less commonly in the distal extremities [1,3,7-10]. Of the distal extremities, lower limb has been found to be more common with 7 reported cases, tibial nerve being most frequently involved [11]. There has been only one reported case involving the upper limb median nerve located proximally in the mid-arm [6]. We describe the first case involving the distal ulnar nerve in the dominant hand of an elderly male and report on his clinical presentation, intra-operative findings, histology and clinical outcome.

Case Report

A 67 year old gentleman presented to us with a 3 year history of gradually increasing painless swelling involving the distal volar aspect of his right forearm. There was no history of any prior trauma and he had no known comorbidities. He also complained of tingling and numbness involving the ipsilateral ring and little finger for the past 3 months. Clinically, he was found to have a well circumscribed 3x2cm firm painless mass involving the flexor aspect of the right distal forearm. The ulna artery was well palpable medial to the mass. Tinel's sign was positive and he had a 30% decrease in sensation in the ulnar nerve dermatome of his hand with no motor deficits. His routine blood investigations were normal. Plain radiograph of the wrist did not show any obvious evidence of any calcification in the tumor region or abnormal ossification of the underlying bone. However, musculoskeletal ultrasound done showed a well circumscribed mass, highly reflective internal hypo echoic areas with calcifications located eccentrically to the nerve axis just abutting the ulnar nerve and artery; a 2.5cm incision was made over the tumor and he underwent marginal resection. Intra-operatively; the mass was found to have a smooth surface, two fascicles were found to be partially adherent to the capsule which was gently and thoroughly dissected out to leave a negative margin and the tumor was freed (**Figure 1**).



Figure 1: Gross intra-operative image showing a well encapsulated smooth surfaced eccentrically located mass, the adherent fascicles were gently freed off the undersurface of the tumor.

Histology revealed a variegated tumour consistency pattern. Grossly the tumors cut section was yellowish with darker tan areas representing hypocellular and degenerative regions. Microscopy showed a diffuse interlacing pattern of hyper and hypocellular cells with presence pleomorphic nuclei in Antoni A areas (**Figures 2a and 2b**).

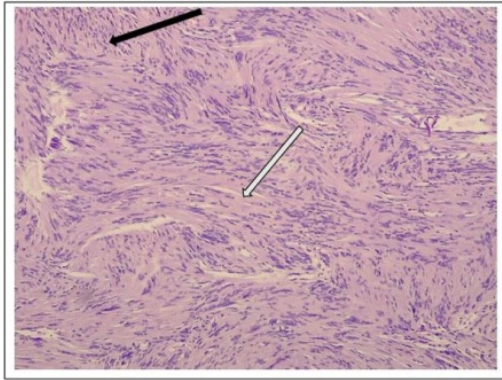


Figure 2a: Low power field showing a biphasic diffuse interlacing pattern of compact hypercellular interlacing fascicles of spindle cells, Antoni A regions (thin black arrow) and loose hypocellular uniform Antoni B areas (thin white arrow).

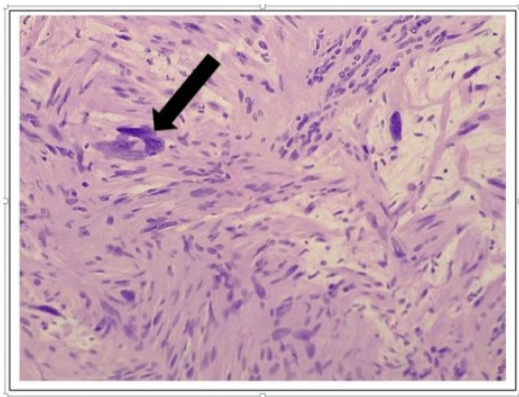


Figure 2b: High power field showing marked nuclear pleomorphism and hyperchromasia (thick black arrow); pathognomic of ancient schwannomma.

The Antoni A cells were spindle shaped with nuclear pleomorphism and had indistinct cytoplasm. They were arranged in thin short bundles along with Verocay bodies. The hypocellular Antoni B areas had a more loosely arranged collagen matrix.

He had an uneventful post-operative period. However, he had mild persisting neuropraxia with sensory deficit involving the ring and little finger which completely resolved over 3 months. At 6 months follow up he was asymptomatic with a well healed surgical scar and had no distal neurovascular deficits (**Figure 3**).



Figure 3: At 6 months follow up:- Well healed surgical scar with no neurological deficits.

Discussion

This is the first case report of an ancient schwannoma involving the ulnar nerve. Late presentations of upper limb schwannomas with distal involvement especially in the dominant hand seems to be fairly uncommon as even a slight neurological compromise can be debilitating, therefore making it less likely for these patients to present late and allow for degenerative changes.

Clinical presentation

Ancient schwannomas have a usual benign indolent long standing course with a rare disposition to malignancy. The common age of presentation is in the elderly. Patient tends to present when they are symptomatic with associated neurological deficits. 1/3rd of these tumors involve the head and neck region, followed by the retro peritoneum, trunk and distal extremities in decreasing order of frequency [3,6,7,11-13]. Of the head and neck region, vestibular and trigeminal involvement is most common, most of these patients tend to present only when symptomatic which occurs over the years with gradual cranial nerve impairment [3,7,10,12,14]. Such is the case with tumors involving the retro peritoneum as well which are more difficult to diagnose till they enlarge in size and cause pressure effects [5,11]. Involvement of the distal extremities is least common. Lower limb involvement has been reported more frequently with 7 cases in literature mainly involving the tibial nerve [11]. Only one case has been reported in the upper limb median nerve [6].

Ours is the first reported case with ulnar nerve involvement. However, the duration of symptoms was only 3 years compared to other reported cases of longer duration, up to even 20 years [11]. The early presentation is probably due to involvement of the dominant hand with sensory compromise.

Radiographic features, surgical findings and histology

Plain radiograph imaging may show calcifications. Ultrasonography reveals hypoechoic areas that correlates with the degenerative cystic changes in a well encapsulated sheath. CT and MRI show enhancement of per degenerative areas near

the capsule. Due to predominance of Antoni B pattern, MRI depicts low signal on T1 and high signal on T2 weighted image. Bone scintigraphy has high uptake, however gallium-67 citrate scintigraphy has no accumulation. These tumors are typically eccentrically located and fascicles can be dissected off from the capsule. The eccentric location of these tumors differentiate it from other peripheral nerve tumors which are commonly located centrally [6]. However infiltration of the fascicles has also been reported and is more common with proximal extremity location, in such cases the individual fascicle along with the sensory or motor supply has to be sacrificed [6]. Diffuse hypo cellular areas with degenerative changes and nuclear atypical is the path gnomonic histological finding.

In our case described, given the age of presentation and location with a characteristic well encapsulated tumor with hypo echoic areas on ultrasonography; further imaging was not carried out. Intra-operatively, the tumor was found to be eccentrically located and the fascicles were not firmly adherent to the capsule as is the case with nerve infiltration in malignant tumors [6]. Both gross and microscopically, the tumor had a characteristic diffuse variegated pattern with a well encapsulated sheath in contrast to a mere reduction of Antoni B cells with cystic areas and nuclear atypical which is commonly described [6,11].

Differential diagnosis

The most common differentials reported are malignant fibrous histiocytoma, malignant peripheral nerve sheath tumor, liposarcoma, synovial sarcoma and haemangiopericytoma [2,11]. Given the long standing tumor, age of presentation and location of these tumors with the head, neck and retro peritoneum most frequently involved, these tumors are often misdiagnosed as malignant on presentation. Few cases of malignant transformation have been previously reported but are still relatively infrequent [15]. As our patient presented with relatively shorter duration of symptoms with clear neurogenic involvement, Tinel sign positive and ultrasound showing high reflection with a visibly free adjacent nerve including internal necrosis, calcification and abnormal vascularity, MRI imaging was not carried out. However routine MRI imaging would be beneficial for craniofacial, trunk and retroperitoneal involvement especially in the absence of neurogenic symptoms. Sudden rapid growth in a long standing tumor, proximally and centrally located tumors along the axis of peripheral nerves with intraoperative infiltration and encasement of fascicles should raise the suspicion of malignancy [4,5,15,16].

The important points that differentiate ancient schwannomas from malignant neoplasms include the age of presentation and long indolent course of the tumors with isolated neurological deficit, clinically positive Tinel's sign, ultrasound showing a visible adjacent nerve abutting the tumor with hyper-reflective echoes, MRI enhancement on T2-weighted images with a well-defined fibrous capsule and split fat sign and hypo cellular areas with nuclear atypical on microscopy [2,11].

Conclusion

Ancient schwannomas are a rarity and diagnosis in the elderly may be challenging. However, keeping in mind the age, long duration, and isolated neurological involvement in the absence of constitutional symptoms along with radiographic and pathognomic histological findings, the diagnosis is difficult to miss. It is interesting to note a unique histologic finding variant in this case seen as a rosette pattern of interlacing variegated cells with nuclear pleomorphism. Early and prompt diagnosis with clinical, radiological, surgical and histological correlation would appear to be the key in getting a good clinical outcome for this rare nerve sheath tumor.

Conflict of Interests

The authors declare that they have no conflict of interest.

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