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Spectrum of schwannomas: No longer a rare entity

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Abstract

Schwannoma is a benign nerve sheath tumour arising from the Schwann cells. Schwannomas are usually solitary in nature. They mimic some of the aggressive and malignant tumours. This study aims to shed some light in the preoperative work-up towards the diagnosis, histopathological highlights, management and the outcomes. This study includes 8 patients over a specific time period with the mean age of 55 years. Although high resolution ultrasound (USG) is beneficial, the Magnetic Resonance Imaging (MRI) is considered the most helpful in the diagnosis that brings out the anatomical relationships and the origin of the tumour. The histopathology of the excised specimens showed the typical characteristics of schwannoma in 6 patients while the remaining 2 patients were diagnosed as ancient schwannoma. Enucleating excision of the tumour from the involved nerve is the preferred treatment option especially under magnification using microsurgical technique. One should be aware of the varied presentation of this pathology depending on the nerve involved and its anatomical location. It is imperative to consider this pathology in the differential diagnosis to avoid any unnecessary and extensive surgical clearance of tumorous swellings.

Keywords: Schwannoma, nerve sheath tumour, benign tumour, tinell's sign, enucleation, antoni A, antoni B

Introduction

Schwannoma is a benign tumour, which develops from the Schwann cells of the nerve sheath. Schwannomas are usually solitary in 95% of cases ^[1], but multiple tumours have also been reported ^[2, 3, 4]. They show a characteristic pattern of slow non-infiltrating growth for several years without any specific symptoms ^[5]. The incidence of schwannomas in the adults and children in eastern countries is 5% and 2% respectively ^[6]. Owing to its low incidence, the clinical signs and symptoms are often misunderstood and are related to other pathologies such as ganglions, tenosynovitis or other soft-tissue tumours ^[7]. The tumour is often associated with a "tingling sensation", but this is not a constant finding at the clinical examination. Hems *et al.* ^[8] reviewed a series of 104 peripheral nerve benign tumours and demonstrated that in only seven cases was the preoperative diagnosis accurate. The surgical enucleation is generally believed to be routinely done for schwannomas without causing neurological deficit as they are well encapsulated ^[9, 10, 11]. This statement does not hold good as reported by other authors ^[12, 13]. This study aims to define the preoperative assessment to help in the diagnosis, and to review the outcomes following the chosen surgical treatment.

Materials and methods

After obtaining ethical clearance from the institutional ethics committee, 8 patients were recruited for the study who presented themselves for treatment between June 2017 and May 2018. Anthropometric and clinical data included age, sex, location of the lesion, symptoms and signs, preoperative investigations, surgical management, histopathological examination, Immunohistochemical staining and the patient follow-up findings. There were 6 male and 2 female patients with the mean age of 55 years (range: 39-72 years) at the time of surgery. The time between the onset of symptoms and the surgical excision ranged from 2 to 5 years. There were 2 schwannomas originating from the posterior tibial nerve, 2 from the superficial peroneal nerve, 1 from the ulnar nerve and the other 3 could not be related to any specific nerve. Topographically, 3 tumours were in the leg, 3 around the ankle region, 1 in the forearm and 1 in the upper arm.

The palpable mass (Figures 1 and 2) was present in all 8 cases of which 6 had peripheral nerve symptoms such as paraesthesia to the territory of the affected nerve or adjacent to the swelling, 3 had a positive Tinel's sign and 6 patients had sensory deficit of varying severity. Two of the 3 patients with tumour around the ankle had tarsal tunnel symptoms for a year. None of the patients presented with any motor weakness. In all the 8 cases, an initial Ultrasound (USG) investigation was performed followed by Magnetic Resonance Imaging (MRI) scan to study the tumour morphology, relationship of the mass with the surrounding structures and the detectability of nerve origin.

Seven patients were treated with surgical enucleating excision of the tumour while one patient underwent excision and sural nerve grafting from the ipsilateral leg. Five patients underwent the procedure under regional anaesthesia while the rest had general anaesthesia. A longitudinal or curvilinear incision was made over the tumour. The tumour capsule was identified and incised to expose the shiny surface of the tumour. With meticulous dissection, the tumour was separated and freed from the surrounding nerve fascicles and was removed in one piece. The intactness of the nerve was checked after tumour excision. Haemostasis was achieved and the wound was closed.

In the patient who underwent sural nerve grafting, the tumour was extensive and was involving more than 50% of the tibial nerve trunk in the tarsal tunnel, before its bifurcation in to the medial and lateral plantar nerves with the medial plantar nerve being majorly involved (Figure 3). With three small incisions along the course of the ipsilateral sural nerve, the nerve was identified, harvested and glued (Figure 4).

All the specimens were sent for histopathological examination and 2 cases sent for immune histochemical staining. Clinical follow-up was at 2 weeks, 6 weeks, 12 weeks, 6 months, one year and then every year. The patients were evaluated for paraesthesia, sensory and motor deficit.

Results

The preoperative diagnosis matched with the postoperative one in 6 out of 8 patients. MRI performed in all these cases suggested schwannoma in 6 cases but could not comprehensively suggest this pathology in the remaining two patients. In spite of the extensive preoperative evaluation, the diagnosis was uncertain in 2 patients and of which in 1 patient the suggested diagnosis was an aggressive pathology thereby requiring an incisional biopsy to clinch the diagnosis before the definitive treatment. The histopathology of the excised specimens showed the typical characteristics of schwannoma in 6 patients (Figure 5) while the remaining 2 patients were diagnosed as ancient schwannoma (Figure 6). The immunohistochemical staining of the two specimens sent, were positive for S100 protein (Figure 7). The complete excision of the mass was possible without much nerve damage in 7 out of 8 patients.

All the patients were reviewed until one year following surgery. Six patients who had varying grades of sensory deficit preoperatively were relieved of this symptom postoperatively except for 1 patient. Postoperatively, Tinel's sign was positive in this same patient who underwent sural nerve grafting along with enucleating excision of the tumour while 2 more patients who had a positive Tinel's sign preoperatively had turned out to be Tinel's sign negative after the surgery. No significant motor deficit was present. There was no recurrence of the tumour until the latest patient follow-up.

Discussion

In the treatment of schwannomas a correct diagnosis is the first essential step^[13, 14]. The preoperative diagnosis becomes difficult due to the risk of misinterpretation as a result of their nonspecific symptoms and a clinical picture which mimics a few other pathology. As reported by Park *et al.*^[14] and Kang *et al.*^[15], the Tinel's sign is the most useful alert sign in the diagnosis. This sign was present in 38% of our patients and this depended on the involved nerve. With an isolated, palpable slow growing mass, a suspected schwannoma should be considered when the mass is located with a positive Tinel's sign usually on the volar surface of the limb^[16]. Although, the diagnosis of a soft tissue tumour is usually clinched by an open biopsy, it is not routinely recommended owing to the high risk of dissemination if the tumour is malignant or for the risk of causing an iatrogenic nerve injury^[14]. Though an ultrasound-guided needle biopsy seems to be a less invasive option, the percentage of success in accurately diagnosing the lesion is less.

Among the diagnostic modalities available pre-operatively, MRI is considered the most helpful in confirming the diagnosis^[8, 13, 14, 15, 16, 17]. MRI brings out the anatomical relationship and the origin of the tumour. The T1 weighted MRI sequences of a schwannoma shows a homogeneous mass, with a low to medium intensity signal while the T2 weighted sequences show a high signal. Due to low intensity signal areas, an ancient schwannoma can produce a different image (Figures 8 and 9), which are caused by hyalin and degeneration^[18, 19]. Ancient schwannoma which has a typical pattern, is an uncommon variant of schwannoma^[16]. This subtype is characterised by extensive degeneration and diffuse hypocellular areas and these changes might occur because it takes a long time for ancient schwannomas to develop^[19]. Ancient schwannomas might be confused with malignant tumours due to their atypical nuclear features and cystic degeneration on histology and imaging, leading to a radical surgical approach.

The 'Split Fat sign' in the MRI helps to differentiate a peripheral nerve tumour (unless malignant) from the others. The 'Target sign' occurs as a result of low intensity signal (fibrous component) in the central portion and a high intensity signal (myxomatous component) in the peripheral part. It has been reported that MRI does not adequately distinguish between schwannomas and neurofibromas^[8, 15]. Schwannomas are frequently encapsulated but neurofibromas are usually not. Detection of a capsule which causes a low intensity rim at the margin of the tumour along one side of the mass with the presence of the nerve on the other side could help to differentiate schwannomas from neurofibromas^[3, 16, 17, 20]. Sometimes, it is difficult to identify the capsule of the tumour with MRI and in such occasions, the ultrasound imaging can be combined with MRI to enhance the precision of pre-operative diagnosis^[19, 21].

In the past, the ultrasound investigation was not able to always reveal the connection between the tumour and the nerve and hence ineffective to make a correct preoperative diagnosis^[22]. With today's advances in ultrasound technology, it can provide higher spatial resolution identifying the location and origin of the nerve tumour and can provide its relationship with other uninvolved nerve fascicles^[23, 24, 25]. As it is extremely operator dependent, a musculoskeletal sonologist is essential to achieve a high rate of diagnostic success.

In our experience, MRI provides high diagnostic value for schwannomas. In the current study, the MRI was able to correctly suggest the pre-operative diagnosis in 75% of our

cases which is superior to those reported by other authors [7, 26], confirming the significance of MRI when approaching nerve tumours [3, 14, 16]. Enucleation of the tumour under magnification using microsurgical technique from the involved nerve is the suggested treatment option [12, 13, 14, 15, 16, 25, 27, 29]. Two main approaches have been reported; extracapsular [15] or intracapsular enucleation [14, 25, 27]. We preferred the intracapsular enucleation due to the lesser risk of neural damage. With such an approach of enucleating excision of the tumour the damage to the nerve fascicles was kept to a minimum and thereby no postoperative residual neural deficit in 7 out of 8 cases. In the remaining case, the direct repair of the nerve was not possible due to the amount of nerve fascicles lost in the resection of the tumour necessitating a nerve graft.

In most cases (82%) the temporary impairment after surgery was present, independent of fascicle preservation. There are mainly three different reasons to explain this findings [13, 28]. Schwannoma starts from the nerve sheath and the containing fascicle is always involved independently from its residual function; the longitudinal incision employed during dissection of the tumour can damage small fascicles; finally, intact fascicles preserved from the tumour growth may be compressed during its surgical enucleation resulting in a neuropraxic injury. The complexity of location, size of the lesion and the 'Nerve of origin' of the tumour can sometimes dictate the final results. In our study, the location of the tumour did not significantly alter the final outcome but none of our patients had schwannomas located in a complex area such as the nerve plexus. Sawada *et al.* [13] reported four cases of schwannoma in the brachial plexus that could not be enucleated completely because of the involvement of many fascicles. In such cases, resection of the tumour performed included some normal fascicles, causing permanent sensory and motor neurological deficits.

In our series, all the schwannomas around the ankle region presented with pain (75% of all painful cases) as the major complaint, whereas only 20% of more proximal lesions were painful. This suggests that pain corresponds to the degree of nerve compression [15, 28], the lack of local tissue distensibility and local pressure effects [7]. Although the schwannoma is cautiously separated from the involved nerve, transient postoperative paraesthesia may still occur. The sensation should recover in a few months to one year in most cases as reported in the literatures [5, 12-16, 25, 28-30]. This is reflected in our study as well, as the complete recovery of sensation was evident in about 88% of cases at 1-year follow-up. Schwannoma has Antoni A and Antoni B areas. The Antoni A area is hypercellular, more organised and is composed of spindle cells arranged in short bundles or interlacing fascicles. The Antoni B area is hypocellular and less organised which contains more myxoid, loosely arranged tissue with a high water content. These areas are intermixed in schwannomas and occur in varying amounts [31].

The clinical presentation of such a schwannoma can sometimes be subtle depending on its anatomical location and especially if the size of the lesion is small. A small sized schwannoma of the posterior tibial nerve could present as a mild tarsal tunnel syndrome with a positive Tinel's sign being the only clinical sign to guide the clinician [32]. Unless there is a high degree of suspicion, the diagnosis can be easily missed at this stage.



Fig 1: Palpable mass on the anterior aspect of lower third of leg (View from the side).



Fig 2: Palpable mass on the anterior aspect of lower third of leg (View from the front).

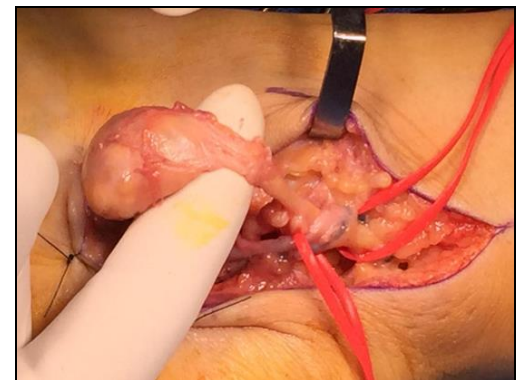


Fig 3: Intraoperative view of schwannoma of the posterior tibial nerve



Fig 4: Intraoperative view after removal of schwannoma from posterior tibial nerve. The sural nerve graft is shown.

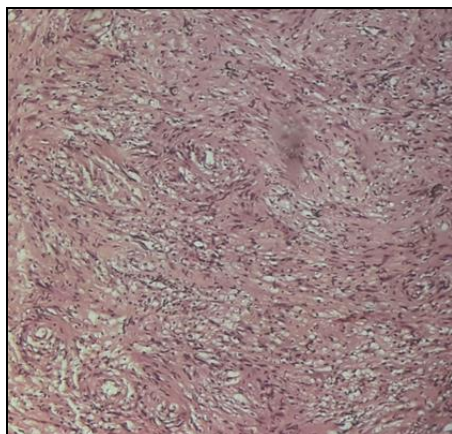


Fig 5: Histopathological slide of schwannoma.

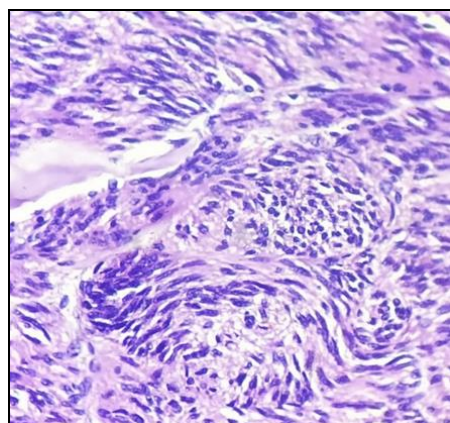


Fig 6: Histopathological slide of ancient schwannoma.

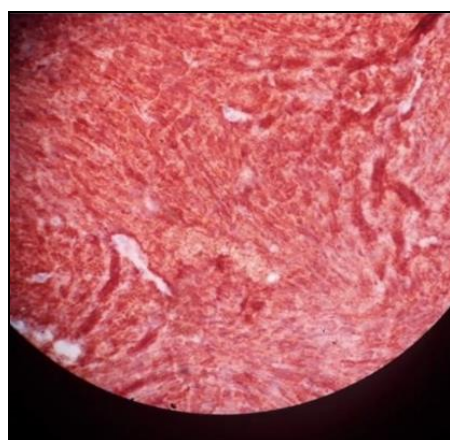


Fig 7: Immunohistochemical staining positive for S100 protein.

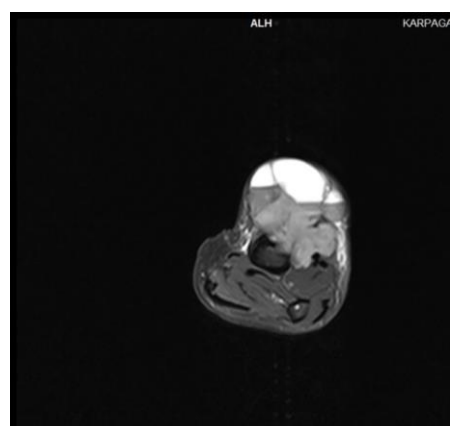


Fig 8: MRI images of ancient schwannoma (transverse plane).



Fig 9: MRI images of ancient schwannoma (sagittal plane).

Conclusion

Schwannoma, a benign nerve sheath tumour is no longer a rare entity. As against the existing myth about the ancient schwannoma with regards to its occurrence in the human body, it is reasonably prevalent in the extremities with an incidence of 25% in our small series. It is imperative to recognise and consider the full spectrum of this pathology in the differential diagnosis as their clinical picture mimics a few 'aggressive treatment demanding' malignant tumours. An integrated assessment using high-resolution USG imaging and MRI is recommended to enhance the diagnostic precision thereby avoiding any unwanted surgeries. This would also help to assess any potential neurological deficits and to plan for any additional procedures at the same time. Albeit the nerve injury risk associated with surgical removal of schwannoma, it is a well-accepted treatment option as the incidence of spontaneous resolution of this iatrogenic neurological deficit is higher, provided a meticulous surgical technique was followed.

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