Giant forefoot schwannoma: case report

Swanoma gigante do antepé: relato de caso

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Keywords:
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ABSTRACT

Benign schwannoma/benign neurilemmoma is a solitary nerve sheath tumor that typically presents in adults between age 20 and 50. Schwannoma accounts for less 5% of benign soft tissue tumors. These tumors may arise in bone as well as in soft tissues. Most lesions are asymptomatic. Schwannoma is rarely found in the foot. The typical solitary tumor presents as a slow growing painless mass which may have been present for 1 to 2 years or more. There can be local bony impingement and bone remodeling because of pressure from the tumor, but these tumors do not invade bone. MRI scans show typical features for an indeterminate tumor, with low signal intensity on T1 and high signal intensity on T2 weighted sequences. Most lesions may be observed without surgery. Because of the indeterminate MRI appearance of this tumor, a complete evaluation and staged biopsy is recommended before definitive surgical removal is planned. Treatment is by excision with a marginal margin. Recurrence following resection is rare. Less than 10% of schwannomas occur in the foot. We report on the presentation and management of a very large schwannoma which caused extensive bone remodeling in the forefoot.

Level of Clinical Evidence: 4

Descritores:
Pé/patologia; Neurilemoma/cirurgia; Relatos de casos

RESUMO

O Schwanoma benigno também conhecido como neurilemoma benigno é um tumor solitário da bainha do nervo tipicamente presente em adultos de 20 a 50 anos de idade. O Schwanoma compreende menos de 5% dos tumores benignos dos tumores de tecidos moles. Podem surgir tanto nos tecidos moles quanto no interior dos ossos. A maioria das lesões são assintomáticas. São lesões raramente encontradas no pé. O tumor típico é solitário e se apresenta como uma massa de crescimento lento e indolor que se pode estar presente por 1 a 2 anos. Pode haver compressão local com remodelação óssea em decorrência da pressão exercida pelo tumor sem que haja invasão óssea. A ressonância magnética (RM) mostra os sinais de um tumor indeterminado com baixo sinal nas sequências em T1 e alto sinal nas sequências em T2. A maioria das lesões pode ser conduzidas sem cirurgia. Em virtude do aspecto indeterminado deste tumor nas imagens da RM a avaliação completa e a biopsia estagiada é recomendada antes do planejamento da remoção cirúrgica definitiva. O tratamento pela excisão cirúrgica deve seguir o padrão de margem segura. A recidiva após a ressecção cirúrgica é rara. Menos de 10% dos schwanomas ocorre no pé. Apresentamos a evolução e tratamento de um schwanoma gigante que causou importante remodelação óssea no antepé.

Nível de Evidência: 4

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INTRODUCTION

Schwannoma is also called neurilemoma, neurinoma, neurocytoma, peripheral glioma and peripheral fibroblastoma.[1,2] World Health Organization Classification of Tumours uses the term schwannoma, which is used here.[3] A schwannoma is a benign, encapsuled and non-invasive tumour that is derived from schwann cells.[4]

Schwannomas are uncommonly found in the foot.[5] The etiology is unknown, but risk factors include trauma and neurofibromatosis type 2.[4] The clinical presentation depends on the location and size of the lesion. The sign and symptoms will typically result from the mass effect and/or direct involvement of the nerve and surrounding tissue.[6]

CASE REPORT

A 69 years old woman with no medical history presented to our clinic with a 9 year history of a mass on the right foot. She had no pain but had difficulty fitting in her shoes. On physical examination a large mass was visible and palpable on the dorsal surface of the lateral forefoot. A larger mass was visible and palpable on the plantar surface of the same location. The borders of the mass were not distinct. There was no pain with palpation. There was no inflammation or redness.

On plain radiographs there was marked splaying of the interspace between the 4th - 5th metatarsals interspace with a bowing deformity of the fourth metatarsal and dramatic thinning of the cortex (Figure 1). Magnetic resonance imaging (MRI) of the foot showed a large mass approximately 6.1 x 4.5 x 3.4cm centered in the plantar aspect of the forefoot. MRI findings were typical for an indeterminate tumor,[7,8] with low signal intensity on T1 and high signal intensity on T2 weighted sequences. The mass had a dumbbell shape with the dorsal portion protruding into the dorsal subcutaneous tissues and splaying the metatarsals (Figure 2).

Large, deep tumors with indeterminate features on MRI scans are very likely to be malignant.[7] As a result this patient required a systematic workup and staged biopsy prior to planning the definitive resection. A complete physical examination revealed no regional or central lymphadenopathy. Laboratory exams were unremarkable. A chest radiograph revealed no mass or nodule. An incisional biopsy was carried out with meticulous attention to hemostasis. The pathological

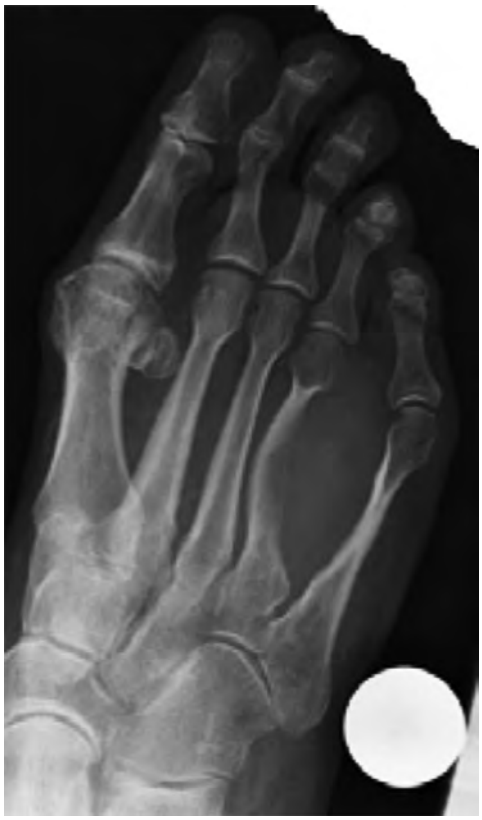

Figure 1. Oblique x ray of the foot, presumed soft tissue mass at the interspace of the 4th and 5th metatarsal bones with associated deformities of these bones. There appears to be a possible involvement of the lateral 4th metatarsal

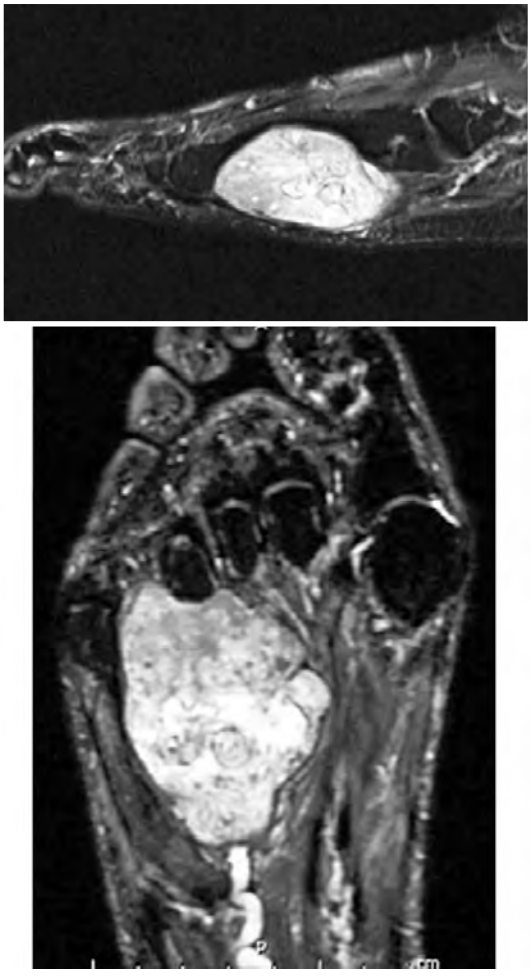

Figure 2. The mass is heterogeneously T2 hyperintense, T1 isointense to muscle, with heterogeneous enhancement. The mass appears encapsulated and displaces the adjacent muscles and tendons, centered in the plantar forefoot displacing adjacent structures with chronic remodeling of the fourth and fifth metatarsals. There is no definite invasion into the surrounding tissues

diagnosis was benign schwannoma with degenerative features (also termed ancient schwannoma).

Surgical procedure

Due to the size of the tumour and its dumbbell shape, two incisions were required. With the patient under general anesthesia in lazy lateral position, a dorsal longitudinal incision was utilized to mobilize the tumor from the superomedial and superolateral sides, and separate it from the metatarsals dorsally. The dissection was intended to result in an uncontaminated marginal margin, which is accomplished by a dissection just outside the capsule of the tumor, with care not to enter or spill the tumour itself. The dorsal extensor tendons were densely adherent to the capsule of the tumour, but most of these were preserved. Then, a second longitudinal incision that made on the plantar surface of the foot. A circumferential dissection was carried out to mobilize the plantar aspect of the tumour and free it from the metatarsals. Once fully mobilized, the dorsal portion of the mass was then pushed down through the metatarsal interspace. The mass was removed *en bloc* through the plantar incision (Figure 3). The overall resection margin was marginal. Routine layered closure was carried out with a closed suction drain. The patient was placed in a short leg splint for 1 weeks and a walking boot for 2 weeks. Full weight bearing in regular shoes was allowed at 3 weeks. The final pathology report demonstrated a large lobulated tumour with a biphasic appearance consistent with benign schwannoma. Antoni A areas of the tumor were very cellular with a spindle shaped cell population showing nuclear palisading and scattered Verocay bodies. Antoni B areas showed less cellular areas of hyalinization, hemosiderin deposition and cystic changes. Nuclear atypia of degenerative nature was present (Figure 4).

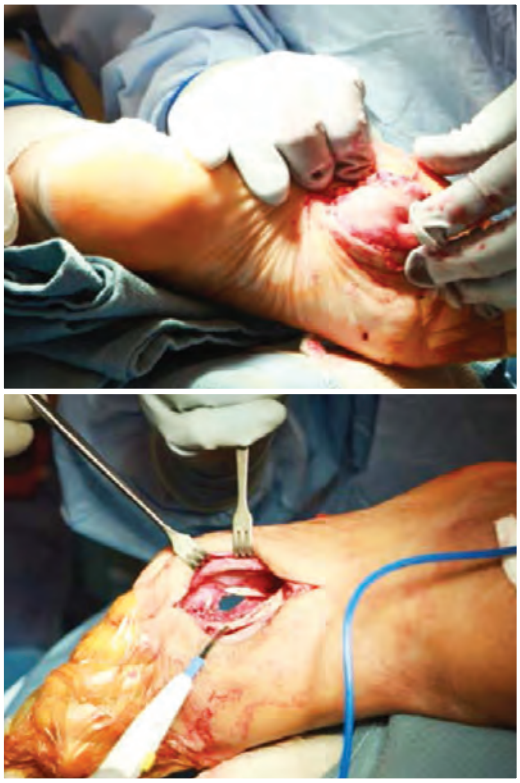

Figure 3. Intraoperative finding, dorsal and plantar approach with deformities of the fourth and fifth metatarsals

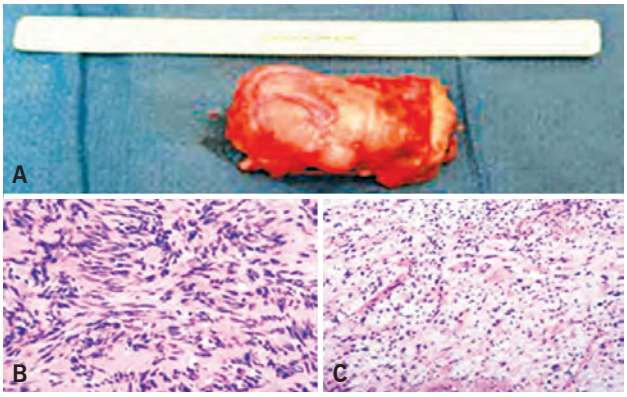

Figura 4. A) excisied mass right foot; B) WS15-7693 10x 2: Medium power Antoni B area, somewhat myxoid hypocellular appearance; C) WS15-7693 20x: Medium-high power Antoni A area, prominent Verocay bodies, nuclear polarization

DISCUSSION

Schwannoma of the foot is interesting due to its rarity. Solitary benign schwannomas have a negligible risk of malignant change which does not appear to be dependent on their size or osseous component. There is no geographical or racial predilection. Schwannoma affects man and women equally with peak age at diagnosis of 30-40 years, and is most commonly localized in the dermis and subcutis with a predilection for head and neck.[9] In one single-center retrospective review, only 10,2% (14 of 137) were located in the foot and/or ankle.[10] In another series, 12 of 104 (11,5%) during 32 years period were located in the foot and ankle.[11] This well encapsulated tumor is usually benign in nature and malignant transformation is rarely reported.[12] However, a careful workup and preliminary biopsy is recommended for all large tumors in the foot. The clinical and imaging features of schwannoma may

be identical to synovial sarcoma, which is the most common malignant sarcoma in the foot.[13] Surgical removal of large tumors in the foot without prior biopsy is not recommended. Preliminary incisional biopsy is safe, it eliminates the potential for a missed cancer diagnosis, and it enables optimal planning of the definitive treatment.[14] Special features of this case are a very slow growing with an unusually long history of 9 year with a long-standing juxtaposed to bone that resulted in bone remodeling at the metatarsal bones. Schwannomas have a good prognosis.[3] The tumours are slow growing and malignant transformation is rare.[2] The discovery of one schwannoma should trigger a careful search for others. This is specially important considering that multiple benign lesions or a malignant schwannoma maybe indicative of other syndromes or pathologies.[12] MRI is particularly useful, it shows a usually round or oval mass with a moderately bright signal on T1-weighted images and a bright, heterogeneous signal on T2-weighted images. The mass is usually less than 2.5cm in size. The lesion enhances uniformly with gadolinium contrast.[12] Early diagnosis and treatment may prevent permanent nerve damage, soft tissue or bony deformity.

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REFERENCES

1. Stout AP, Carson W. The pepipheral manisfestations of the specific nerbe sheath tumours (neurilemoma). Am J Cancer. 1935;24:751-89.
2. Rockwell GM, Thoma A. Schwannoma of the hand and wrist. Plast Reconstr Surg. 2003;111(3):1227-32.
3. Fletcher, C. Unni, K. Mertens, F. World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyaon: IARC Press; 2002.
4. Ferner RE, O'Doherty MJ. Neurobibroma and schwannoma. Curr Opin Neurol. 2002;15(6):679-84.
5. Still G. Neurilemoma of the medial plantar nerve: a case report. J Foot Ankle Surg.2001;40(4):236-9.
6. Mrugala M, Batchelort, Plotkin S. Peripheral and cranial nerve sheath tumors. Curr Opin Neurol. 2005;18(5):604-10.
7. Jong B, Shahabpour M, Spruyt D, et Al. Imaging and differential diagnosis of synovial sarcoma. J Belge Radiol. 1992;75(4):335-9. PMID: 1334066.
8. Ghaly RF. A posterior tibial nerve neurilomoma unrecognized for ten years: case report. Neurosurgery 2001;48(3):668-72.
9. Datir A, James SL, Ali K, Lee J, Ahmad M, Saifuddin A. MRI of soft-tissue masses: the relationship between lesion size, depth, and diagnosis. Clin Radiol. 2008;63(4):373-8; discussion 379-80.
10. Wu J, Hochman M. Soft-tissue tumors and tumorlike lesions: a systematic imaging approach. Radiology. 2009;253(2):297-316.
11. Iwashita T, Enjoji M. Plexiform neurilemmoma: a clinicopathological and immunohistochemical analysis of 23 tumours from 20 patients. Virchows Arch A Pathol Anat Histopathol. 1987;411(4): 305-9.
12. Ruggieri M. The different forms of neurofibromatosis. Child Nerv Syst. 1999;15:295.
13. Fortnum H, O'Neill C, Taylor R, Lenthall R, Nikolopoulos T, Lightfoot G, et al. The role of magnetic resonance imaging in the identification of suspected acoustic neuroma: a systematic review of clinical and cost effectiveness and natural history. Health Technol Assess. 2009;13(18):iii-iv, ix-xi, 1-154.
14. Kehoe NJ, Reid RP, Semple JC. Solitary benign peripheral-nerve tumours: review of 32 years experience. J Bone Joint Surg Br. 1995;77(3):497-500.
15. Harkin JC, Reed RJ. Tumors of the peripheral nervous system, fascicle 3, second series. Washington, DC: Armed Forces Institute of Pathology; 1969. p.60-4.