

SUBPERIOSTEAL SCHWANNOMA OF THE HUMERUS: A CASE REPORT AND REVIEW OF LITERATURE



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ABSTRACT

Numerous conditions arise from the surface of bones, including both neoplastic and non-neoplastic lesions. Schwannoma had been reported to be one of those conditions originating from the bone surface with variable proposed theories on its origin. We present a unique case of juxtacortical schwannoma originating from the humerus in a 40-years-old female with a clinical presentation of painful swelling for two years. The Radiograph showed a cortically-based lytic lesion in the lateral distal humeral metaphysis with cortical scalloping. Magnetic resonance imaging showed the same juxtacortical lesion with an extraosseous soft tissue component. It demonstrates intermediate T1 and high T2 signal intensity with intense enhancement. Ultrasound-guided biopsy was performed, and histopathology findings showed a benign peripheral nerve sheath tumour consistent with schwannoma. The patient underwent complete resection with no complications or documented recurrence till the latest follow-up. In conclusion, we present a case of a pathology-proved juxtacortical schwannoma in a patient who has been experiencing vague painful swelling for a prolonged period of time and we review and summarize the previously reported cases.

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خلاصة

تنشأ العديد من الحالات المرضية من سطح العظام بأشكال ورمية وغير ورمية. ذكرت الأبحاث السابقة الورم الشفاني كأحد تلك الحالات التي تنشأ من سطح العظم وفق نظريات متعددة اختلفت حول أصله. نقدم حالة فريدة من الورم الشفاني المجاور للقشرة التي نشأت من عظم العضد لدى أنثى تبلغ من العمر 40 عاماً تشتمل على تورم مؤلم لمدة عامين. أظهر التصوير بالأشعة السينية آفة بقشرة عظم الكردوس العضدي في الجزء الجانبي القاصي مع وجود توقع قشري. كما أظهر التصوير بالرنين المغناطيسي نفس الآفة مكونة من أنسجة رخوة خارج العظم. حيث ظهرت بكثافة في إشارة T1 المتوسطة وكثافة عالية في إشارة T2. تم إجراء خزعة تحت توجيه الموجات فوق الصوتية وأظهرت نتائج تحليل الأنسجة وجود ورم حميد في غمد العصب المحيطي يتوافق مع الورم الشفاني. خضع المريض لاستئصال كامل دون مضاعفات أو تكرار مؤثق حتى آخر متابعة. في الختام، يقدم هذا التقرير حالة من الورم الشفاني المجاور لدى مريض عانى من تورم مؤلم غامض لفترة طويلة من الوقت، ونراجع الدراسات السابقة ذات العلاقة بهذه الحالة.



1. INTRODUCTION

Schwannomas are encapsulated slow growing benign tumours originating from the neural crest cells of the peripheral nerve sheath called Schwann cells [1, 2]. The main function of these cells is to produce the myelin sheaths surrounding peripheral nerves [1]. Schwannomas are rare and usually involve the soft tissues of the head and neck [3]. However, occasionally it may arise from the bone either within the medullary cavity of the bone, known as intraosseous schwannoma most commonly arising from the mandibular nerve or in extremely rare cases, originate from the surface of the bone or from the parosteal tissues, also known as subperiosteal or juxtacortical schwannoma [1, 4, 5]. To our knowledge, only 7 case reports and 1 case series are present in the literature describing bone surface schwannomas. We present a case of juxtacortical schwannoma originating from the posterior surface of the humerus, discuss the suggested theories on the origin of bone surface schwannomas and summarize prior case reports.

2. CASE PRESENTATION

A 40 years old female patient, otherwise healthy, was referred to the sarcoma clinic for further evaluation and management of a painful swelling at the left elbow for two years. There was no history of trauma reported before. Clinical examinations showed elbow swelling laterally, tender on palpation, normal range of motion of the elbow and intact neurovascular examination. There were no other similar lesions, and no café-au lait spots or cutaneous neurofibromas. Elbow radiograph showed a cortically based lytic lesion located in the lateral distal humeral metaphysis with cortical scalloping (Figure 1).



Figure 1 A-Frontal and B-lateral elbow radiographs showing: cortical based lytic lesion in distal humeral metaphysis associated with and cortical scalloping.

MRI showed an oblong juxtacortical lesion along the posterolateral cortex of the distal humeral metaphysis causing cortical scalloping with an extraosseous soft tissue component resulting in thinning and elevation of the periosteum. The mass demonstrates homogenous intermediate T1 signal intensity and high signal intensity on T2 weighted images with intense enhancement in the post contrast sequences (Figure 2). There were no calcifications, no cystic changes or blooming artifact.

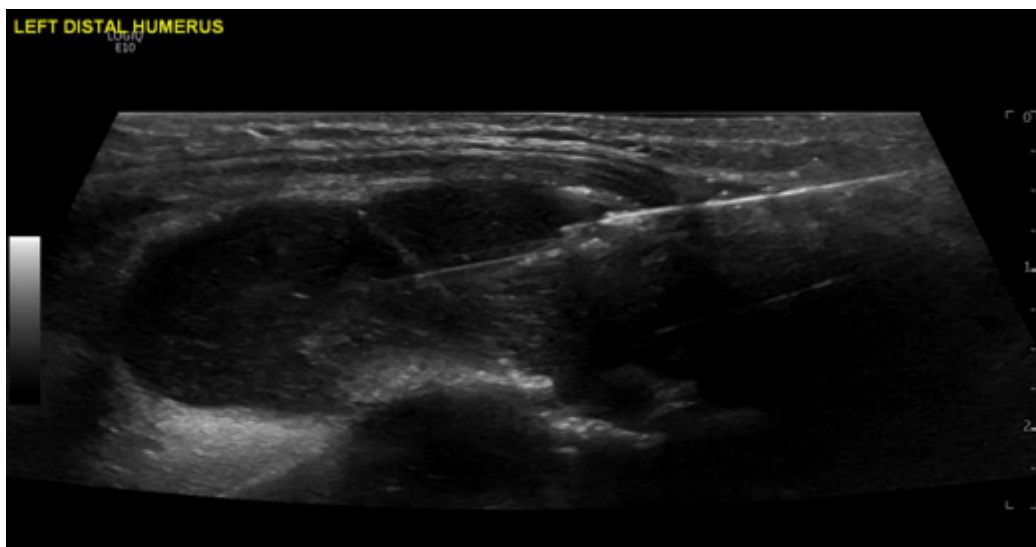


Figure 2 Axial A. T1 Fat sat, B. T2 Fat sat, C. T1 Fat sat post contrast, D. T1 non-Fat sat and E. Sagittal T2 Fat sat MRI images showing: oblong juxtacortical lesion along the posterolateral cortex of the distal humeral metaphysis causing cortical scalloping with an extraosseous soft tissue component resulting in thinning and elevation of the adjacent periosteum. The lesion demonstrates homogenous intermediate T1 and high T2 signal intensity within tense enhancement in the post contrast images.

Since a majority of the mass was within the soft tissues and palpable by examination, ultrasound guided biopsy was done (Figure 3). Histopathology results showed benign peripheral nerve sheath tumour, consistent with schwannoma. No necrosis or increased mitoses were seen (Figure 4 A-C). Immunohistochemistry staining demonstrated diffusely strong positive S100 staining (Figure 4 D). Eventually, the patient underwent marginal resection. During surgery the mass was attached to the surface of the bone and was following the course posterior cutaneous nerve which was the most likely site of origin. The tumor was excised entirely and the nerve was intact. There were no immediate or delayed complications. There is no documented recurrence till the latest follow-up.

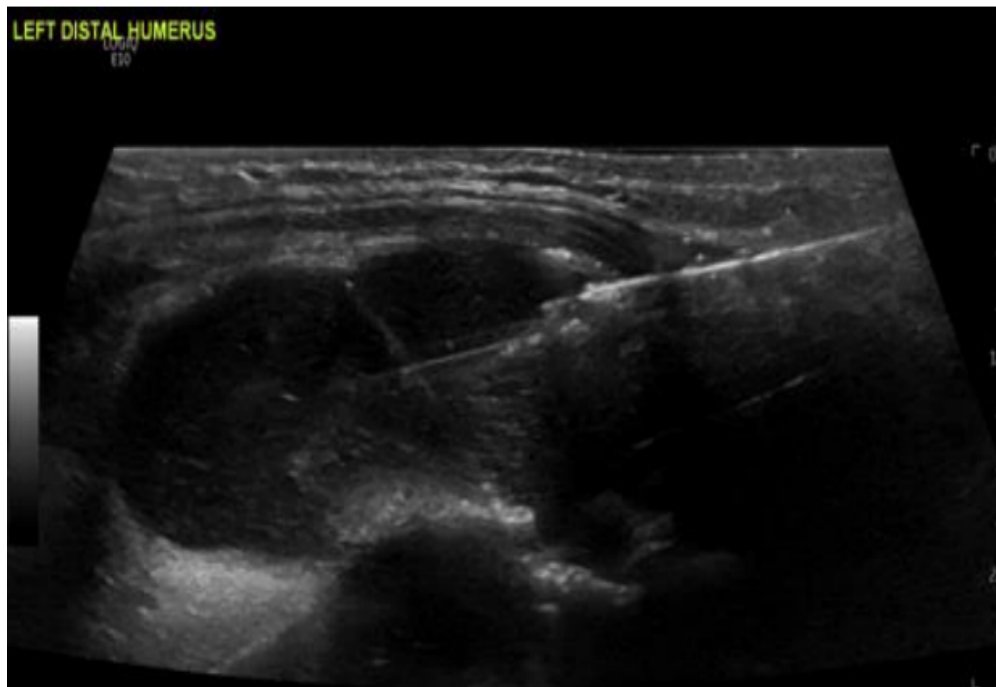


Figure 3 Image from ultrasound-guided biopsy shows the coaxial biopsy needle within a well-defined lobulated hypoechoic lesion next to area of irregular bone cortex.

3. DISCUSSION

Surface bone lesions are a category of tumors with extramedullary origin regardless of their exact anatomical relation to the periosteum [6]. This may include tissues from the inner surface of the cortex to those external to the periosteum [7]. Many neoplastic and non-neoplastic conditions can originate from the surface of the bone, some of these lesions have a characteristic imaging appearance. In contrast, many other conditions show non-specific findings, which can result in a diagnostic challenge [6, 7].

Schwannoma is typically a benign encapsulated lesion that can arise from any peripheral nerve, including those nerves supplying the periosteum; therefore, it may be a potential differential consideration of juxtacortical lesions [8].

Several proposed theories exist in the literature on the origin of bone surface schwannomas. The first theory proposes these tumours develop from traversing nerves across the osseous canal or the nerves accompanying the nutrient vessels supplying the periosteum. The second theory is that it primarily arise in the surrounding soft tissues, causing secondary erosions and extending to the bone surface appearing as juxtacortical lesions [9–11]. The latter theory is supported by the findings in our case as the mass most likely originated from the posterior cutaneous nerve. Nonetheless, it remains difficult to explain why bone surface schwannomas are so rare, as the periosteum has numerous nerve supply.

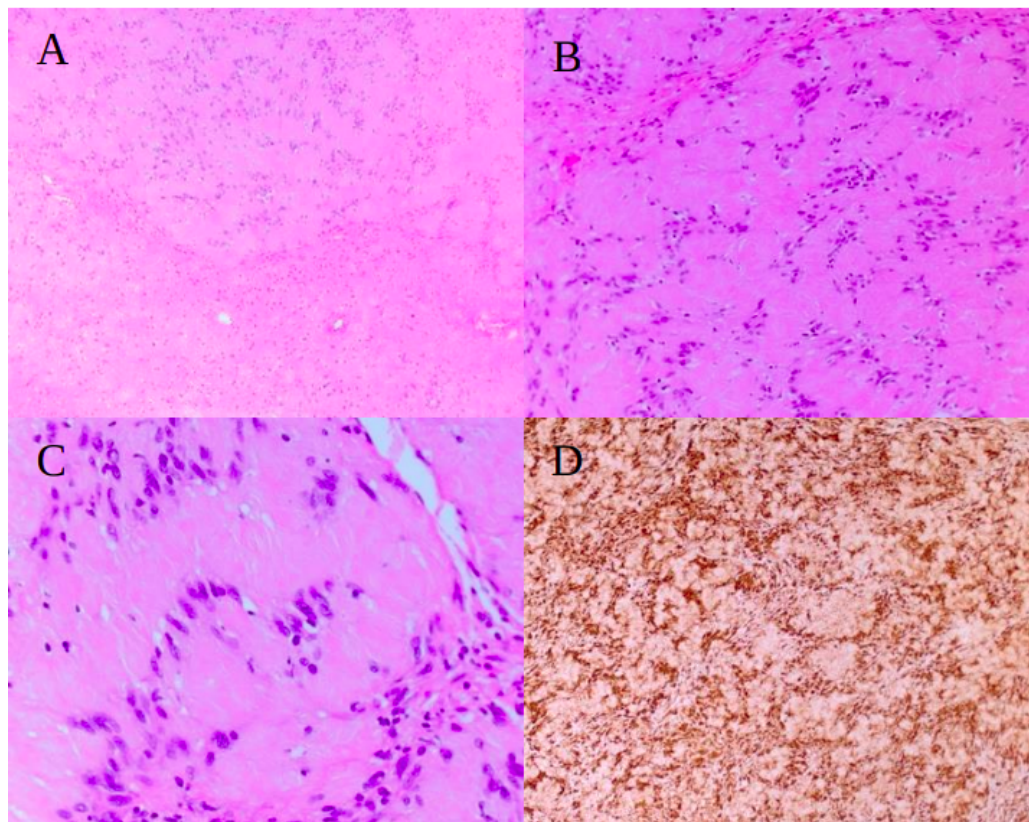


Figure 4 A-C. H&E stain shows spindle cell neoplasm composed of biphasic areas with hypercellular and myxoid hypocellular areas with prominent nuclear palisading around fibrillary processes (Verocay bodies). No necrosis or increased mitoses. D- S100 immunostain is diffusely and strongly positive.

The first case of juxtacortical schwannoma was reported in 1993 by Andrew S.M. et al. in a 52 years old male who presented with enlarging thigh swelling. The patient was diagnosed with malignant juxtacortical schwannoma and treated by amputation [5]. Afterwards, there were six more case reports of juxtacortical schwannoma and only one case series (Table 1). Painful swelling was the most common clinical presentation, including the present case [5, 12–15]. On the other hand, age was quite variable amongst the reported cases since there are cases reported in the age group ranging between 18–66 years. The femur was involved in 5 cases [4, 5, 12, 16, 17], while the tibia was involved in three cases [12, 15]. Other involved sites included the pelvis [13], ulna [14] and humerus in the present case.

Imaging is an essential diagnostic tool for evaluating patients with suspected juxtacortical lesions. The majority of these reported cases were manifested in radiographs as cortically based lytic lesions [13–16]; two cases showed soft tissue swelling with periosteal reaction on radiographs [5, 17]. However, there was the exception of the three cases described by Patro BP et al. in their case series having completely normal radiographs despite the long

standing symptoms [12]. On MRI, the tumour appeared as juxtacortical bone lesion with isointense/low signal intensity on T1 weighted images and hyperintense signal intensity on T2 weighted images with two described enhancement patterns, including: homogenous enhancement [16] and target pattern enhancement [4].

All reported cases of juxtacortical schwannomas, including ours, were treated with complete surgical excision and had favourable outcomes. However, there remains a lack in the data about the recurrence rate.

4. CONCLUSION

A correct pre-operative diagnosis of juxtacortical lesions can be difficult. Schwannomas can be included as a potential differential diagnosis for surface lesions of the bone associated with unexplained vague pain and long standing swelling. In our case, the lesion was located with proper clinical examination and radiological imaging, although the definitive diagnosis was made only after biopsy and histopathology examination. Such lesions in a subperiosteal location should be excised entirely to minimize the possibility of recurrence or malignant transformation.

Table 1 Summary and comparison of case reports of subperiosteal schwannoma.

Author	Age	Location	Clinical presentation	Radiograph	MRI
Andrew SM et al. [5]	52	Femur	Painful swelling	Soft tissue swelling with periosteal reaction.	-
	34	Femur	Pain		
	30	Tibia	Painful swelling		
	45	Tibia	Painful swelling		
Al-Lhedan et al. [16]	18	Femur	Swelling	Cortically based lytic lesion.	Cortically based mass with exophytic T1 hypointense, T2 hyperintense soft tissue mass with homogenous enhancement.

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Table 1 continued

Verma et al. [4]	38	Femur	Pain	Cortical scalloping.	Two soft tissue masses, T1 isointense and hyperintense on fluid sensitive sequence with target pattern of enhancement associated with cortical erosions.
Julfiqar et al. [17]	23	Femur	Swelling	Soft tissue swelling with periosteal reaction.	Lobulated juxtacortical heterogeneous mass with periosteal elevation.
Lakothia D et al. [13]	34	Pelvis	Painful swelling	Cortically based lytic lesion.	Two cystic lesions, T1 isointense T2 hyperintense.
Vivek et al. [14]	28	Ulna	Painful swelling	Cortically based lytic lesion.	Juxtacortical cystic lesion.
Moussa M K et al. [15]	66	Tibia	Painful swelling	Cortically based lytic lesion.	Juxtacortical lesion, isointense on T1 and hyperintense on T2.
Present Case	40	Humerus	Painful swelling	Cortically based lytic lesion with periosteal reaction and cortical scalloping.	Juxtacortical lesion causing cortical scalloping with extraosseous soft tissue component. It demonstrates T1 isointense signal intensity and high signal intensity on T2 with homogenous enhancement.

CONFLICT OF INTEREST

There is no conflict of interest to declare

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N/A

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