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CASE REPORT

Synovial Haemangioma of the Elbow

A rare paediatric case and imaging dilemma

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ورم وعائي دموي زليلي في المرفق

حالة نادرة في طفل، ومعضلة تشخيص إشعاعي

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ABSTRACT: Synovial haemangiomas are rare benign vascular proliferations arising in synovium-lined surfaces. While the knee is by far the joint most commonly involved, this condition can also occur in the elbow. We report an eight-year-old boy who presented to the National University of Malaysia Medical Centre, Kuala Lumpur, Malaysia, in 2016 with a left elbow swelling of one year's duration. Magnetic resonance imaging showed a lobulated intra-articular mass with intermediate signal intensity on T1-weighted imaging and low signal punctate and linear structures within the hyperintense mass on T2-weighted imaging. In addition, there was heterogeneous yet avid contrast enhancement on post-gadolinium contrast images. The mass had juxta-articular extension and bony erosion to the coronoid process and the head of the *radius*. Synovial haemangiomas present a diagnostic dilemma. This report highlights certain imaging characteristics to distinguish this entity from other differential diagnoses.

Keywords: Synovial Membrane; Hemangioma; Sarcoma; Magnetic Resonance Imaging; Diagnostic Imaging; Case Report; Malaysia.

الملخص: تعد الأورام الوعائية انتشارات وعائية حميدة ونادرة الحدوث تنشأ من أسطح الغشاء الزليلي. وتحدث هذه الحالات في الغالب في الركبة، إلا أنها قد تحدث أيضا في المرفق. نعرض هنا لحالة طفل عمره ثماني سنوات أحضر إلى المركز الطبي لجامعة ماليزيا الوطنية في كوالا لامبور بماليزيا في عام 2016. وكان ذلك الطفل مصابا بورم في مرفقه الأيسر منذ عام كامل. وتبين من تصوير الرنين المغنطيسي وجود كتلة مفصصة بين المفاصل كانت لها إشارة متوسطة الكثافة على تسلسل التصوير في T1 المرجحة، وإشارة قليلة الكثافة مُنقَّطة البنيات الخطية في داخل الكتلة عالية الكثافة بتسلسل التصوير في T2 المرجحة. وبالإضافة لذلك كان هنالك تضخيم متغاير وشديد الحدة للتباين في صور ما بعد عنصر الغادولينيوم. وكان للكتلة امتداد مجاور للمفصل، وتآكل للعظم في النَّاتِئُ الإِكْلِيلَانِيُ ورَأْسُ الكَعْبُرَة. تمثل الأورام الوعائية الدموية الزلالية معضلة تشخيصية. ويبرز تقرير الحالة هذه بعض خواص الصور التي تميزَ هذه الحالة من غيرها بالتشخيص التفريقي.

الكلمات المفتاحية: غشاء زليلي؛ ورم وعائي دموي؛ ساركوما؛ تصوير الرنين المغنطيسي؛ التصوير التشخيصي؛ تقرير حالة؛ ماليزيا.

Synovial haemangiomas are rare benign vascular lesions first described by Bouchut in 1856.[1] Most cases involve young adults and children; moreover, men are affected more often than women.[2] Due to their potential for significant morbidity, synovial haemangiomas require early diagnosis and treatment.[2,3] This case report describes the clinical presentation, differential diagnosis and imaging findings of a histologically-proven case of synovial haemangioma of the elbow.

Case Report

An eight-year-old boy presented to the National University of Malaysia Medical Centre, Kuala Lumpur, Malaysia, in 2016 with a swelling in his left elbow that his parents had first noticed one year prior. There was no history of antecedent trauma to the affected elbow. The patient had been previously seen by a general practitioner who treated the swelling as an infection and prescribed a course of antibiotics. Due to a lack of improvement following completion of the antibiotic course, the patient was referred to an orthopaedic surgeon who prescribed a course of non-steroidal anti-inflammatory drugs to which he responded positively. At this stage, the probable working diagnosis was of an osteoid osteoma. Upon physical examination, there was fixed flexion of the elbow to approximately five degrees. The mass measured 5 x 3 cm and was oval, soft, non-tender, immobile, not warm and had a smooth surface with ill-defined margins.

A plain radiograph of the left elbow showed a mass or effusion causing signs of a posterior fat pad and anterior sail [Figure 1]. No abnormal calcification was seen. In view of the persistent swelling, a magnetic resonance imaging (MRI) examination was performed. The MRI scans showed an ill-defined multilobulated intra-articular lesion with an epicentre proximal to

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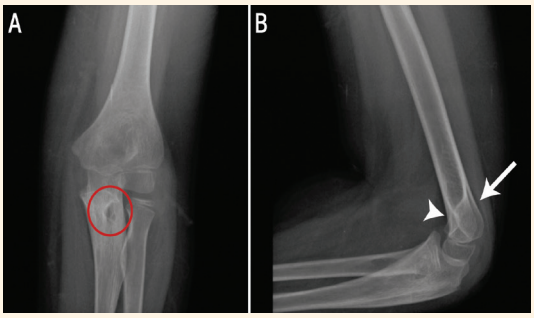

Figure 1: Plain radiographs of the left elbow of an eight year-old boy from the **(A)** anteroposterior and **(B)** lateral views. Signs of a posterior fat pad (arrow) and anterior sail (arrowhead) indicated an effusion or mass in the left elbow joint. A lucent/lytic lesion (circle) was seen on the proximal left *ulna*, potentially representing a cortical bony erosion.

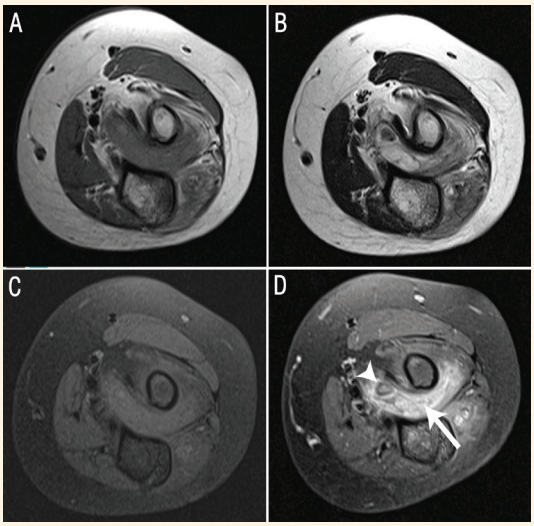

Figure 2: Magnetic resonance imaging scans of the left elbow of an eight year-old boy from the axial view showing an ill-defined and multilobulated lesion, including **(A)** T1-weighted, **(B)** T2-weighted, **(C)** T1-weighted fat-saturated and **(D)** T1-weighted post-gadolinium contrast images. The epicentre of the mass was located in the proximal intramedullary region involving the *supinator* muscle in the posteromedial compartment of the left proximal forearm. The lesion was slightly hyperintense on T1-weighted imaging and hyperintense on T2-weighted imaging, with internal linear and punctate hypodensities. The signal intensity was not suppressed on fat saturation. Post-gadolinium administration, the lesion demonstrated avid yet heterogeneous enhancement (arrow) with a well-defined oval calcification (arrowhead).

the intramedullary region of the left elbow with extra-articular extension. It involved the *supinator* muscle in the posteromedial compartment of the left proximal forearm. It was slightly hyperintense on T1-weighted imaging (T1WI) and hyperintense on T2-weighted imaging (T2WI), with internal serpentine linear and punctate hypodensities [Figures 2A and B]. The signal intensity was not suppressed upon fat saturation

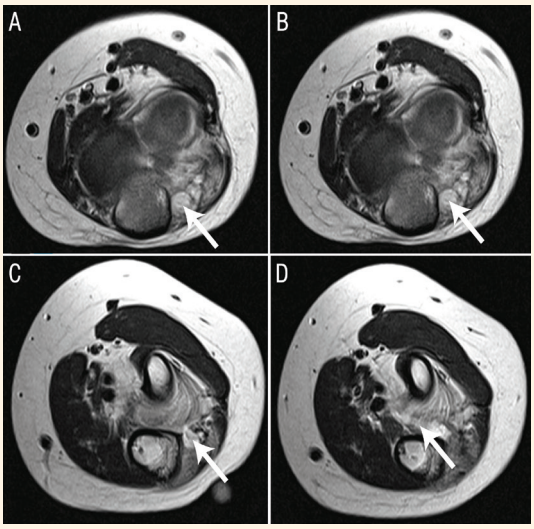

Figure 3: T2-weighted magnetic resonance imaging scans of the left elbow of an eight year-old boy in the superior to inferior positions. A hyperintense tubular structure could be seen arising from a lobulated mass in the posteromedial part of the left elbow and crossing the proximal left *ulna* anteriorly to the anterior compartment of the forearm (arrows).

[Figure 2C]. Post-gadolinium administration, the mass demonstrated avid yet heterogeneous enhancement [Figure 2D].

Further T2WI scans indicated the presence of a tubular-like hyperintense structure within the lesion, potentially representing a vascular component [Figure 3]. A cortical breach into the intramedullary region at the level of the coronoid process of the *ulna* and into the proximal *radius* shaft was also noted. The lesion extended posteriorly to infiltrate the *anconeus* muscle and was relatively large in comparison to the cortical breach [Figures 4A–C]. A core biopsy confirmed features of a benign vascular neoplasm, with a final diagnosis of a synovial haemangioma [Figure 4D]. There was no evidence of calcification, atypical cells, multinucleated giant cells, granulomas or malignancy. An ultrasound showed the lesion to be a diffuse hypervascular mass. Since there was no worsening of the symptoms, the parents of the patient opted for follow-up imaging, after being informed of the risks and benefits of resection surgery.

Discussion

While most synovial haemangiomas tend to involve the knee joint, this type of lesion can occur in any joint of the body, including the intra-articular space, ankle, temporomandibular joint and *bursae*.[2–5] Devaney *et al.* reported 20 cases of synovial haemangioma, of which only six had originated from within the elbow.[2] Such patients can present with multiple symptoms,

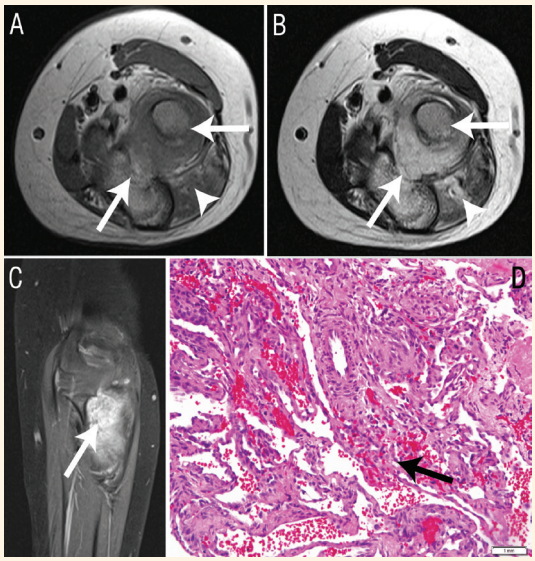

Figure 4: A & B: T1-weighted magnetic resonance imaging (MRI) scan in the axial view and T2-weighted scan in the coronal view of the left elbow of an eight year-old boy showing a cortical breach into the intramedullary region at the level of the coronoid process into the proximal *radius* shaft (arrows) and extending posteriorly to infiltrate the *anconeus* muscle (arrowheads). **C:** T1-weighted MRI scan in the coronal view showing the large size of the lesion (arrow) compared to the cortical breach. **D:** Haematoxylin and eosin stain at x20 magnification showing predominantly fibrous tissue, adipose tissue and skeletal muscle fibres. Note the numerous admixtures of dilated vascular spaces interspersed between the connective tissue matrix (arrow) consisting of both thick-walled, thin-walled and capillary-sized vessels lined by bland-looking endothelial cells.

including persistent elbow pain, swelling, loss of range of movement either in terms of flexion or extension and posterior interosseous nerve palsy secondary to a haemangioma.[6–9] Despite its rarity, the early diagnosis of a synovial haemangioma is important as recurrent haemarthrosis may lead to irreversible joint damage and chronic inflammatory synovitis.[10] Chronic synovial haemangioma can also result in pressure erosion or remodelling of the adjacent bones.[11]

The characteristic MRI features of a synovial haemangioma include homogeneous, low or iso-intense signals upon T1WI.[11,12] However, with T2WI, synovial haemangiomas have lobulated, well-defined margins with heterogeneous high signal intensity. Moreover, multiple low signal punctates or linear structures may be present within the lesion.[11,12] Following the administration of gadolinium, synovial haemangiomas usually demonstrate heterogeneous enhancement. Magnetic resonance angiography is a suitable noninvasive imaging method that can be used to evaluate the relationship of the haemangioma to adjacent feeding vessels for better surgical planning.[11,12] Conventional radiography such as extremity X-rays is usually performed in more than half of synovial haemangioma cases.[7] Radiographs commonly show a non-specific soft tissue mass. Localised phleboliths may also be seen and are fairly specific indications of a haemangioma.[7]

The precise diagnosis of synovial tumours is often difficult due to the non-specific nature of the patient's clinical symptoms and physical examination findings. The differential diagnoses of an elbow mass include synovial sarcomas, synovial haemangiomas, pigmented villonodular synovitis (PVNS), synovial chondromatosis, chronic infections of mycobacterial origin, inflammatory arthritis with *pannus* formation, ganglion cysts or lipoma *arborescens*.[7] In the present case, the lesion was unlikely to be PVNS as the condition usually demonstrates low signals on all sequences as a result of a 'blooming' phenomenon in gradient echo sequences.[11,12] Moreover, even though the patient presented with an intra-articular nodular ossified body, synovial chondromatosis usually occurs in adults rather than in young children; moreover, juvenile rheumatoid arthritis is rare before the age of 10 years.[7] A chronic mycobacterial infection was also considered improbable as there was no evidence of necrosis or marrow oedema.[7]

Ganglion cysts demonstrate a low signal intensity on T1WI and a high signal intensity on T2WI; in addition, low signal intensity structures may be present within the lesion.[13] However, this was not considered in the differential diagnosis for the current case as ganglion cysts usually have peripheral rim enhancement, which was not evident. Lipoma *arborescens* is a monoarticular condition that most commonly occurs in the suprapatellar recess and the knee joint.[13] It has frond-like synovial projections of fat signal intensity on MRI, which were not seen in the present case, and rarely has bony erosions. Despite the use of gadolinium contrast, differentiating synovial sarcomas and haemangiomas on radiology images can be challenging. In particular, the characteristic MRI features of a synovial haemangioma may coincide with those of malignant lesions such as intra-articular synovial sarcomas.[12,13] However, synovial sarcomas are associated with calcification in one-third of cases and are not commonly encountered in children. Moreover, although synovial sarcomas arise near the joint, it is rare for them to originate or extend intra-articularly.[13] In the present case, the epicentre of the mass arose from the elbow joint with juxta-articular extension, lowering the likelihood of a synovial sarcoma.

In the current case, there was a cortical breach of the coronoid process of the left *ulna* and proximal *radius*. This therefore made it challenging to determine whether the bony erosion was due to the presence of

a bone tumour which had infiltrated into the adjacent soft tissue or because of a soft tissue tumour that had eroded the adjacent bone. In this case, as the soft tissue lesion was much larger than the cortical erosion, a primary soft tissue abnormality with secondary involvement of the bone was deemed more probable. It is important to note that although the radiographical and MRI features of the present case pointed towards the diagnosis of a synovial haemangioma, it is a rare disease entity and some imaging features may coincide with other differential diagnoses as described above. Therefore, while the presence of the aforementioned imaging features indicate a likely diagnosis of a synovial haemangioma, a definitive diagnosis requires histopathological confirmation via biopsy, as in the present case.

The treatment of synovial haemangiomas depends on the extent of the lesion and should therefore be individualised. Currently, the gold standard of treatment is surgery. Arthroscopic resection may be performed in lesions that are pedunculated or well-defined.[14] If the synovial haemangioma is large and involves intra- and extra-articular regions, then removal via open surgery with local excision or a total synovectomy is preferable. Recurrence rates after surgical removal are higher for lesions with diffuse involvement.[14]

Conclusion

Synovial haemangiomas are rare tumours requiring early diagnosis and treatment to prevent long-term morbidity. However, the diagnosis is challenging due to a myriad of similar conditions. Currently, an accurate diagnosis depends upon a combination of MRI scans, early arthroscopic assessment and histopathological confirmation following a core biopsy. Both clinicians and radiologists should consider this entity in the differential diagnosis of patients who present with synovial lesions.

References

1. Bouchut ME. [Erectile tumor of the knee joint]. Gaz Hop (Paris) 1856; 29:379.
2. Devaney K, Vinh TN, Sweet DE. Synovial hemangioma: A report of 20 cases with differential diagnostic considerations. Hum Pathol 1993; 24:737–45. doi: 10.1016/0046-8177(93)90010-E.
3. McNeill TW, Ray RD. Hemangioma of the extremities: Review of 35 cases. Clin Orthop Relat Res 1974; 101:154–66.
4. Wen DW, Tan TJ, Rasheed S. Synovial haemangioma of the knee joint: An unusual cause of knee pain in 14-month old girl. Skeletal Radiol 2016; 45:827–31. doi: 10.1007/s00256-016-2356-0.
5. Atkinson TJ, Wolf S, Anavi Y, Wesley R. Synovial hemangioma of the temporomandibular joint: Report of a case and review of the literature. J Oral Maxillofac Surg 1988; 46:804–8. doi: 10.1016/0278-2391(88)90195-4.
6. Mohammadi A, Rosa M, Wolfson D. Synovial hemangioma of the elbow: An uncommon lesion to be considered. Internet J Pathol 2008; 7:3.
7. Chatha DS, Rybak LD, Wittig JC, Desai P. Orthopaedic-radiology-pathology conference: Elbow mass in a 9-year-old girl. Clin Orthop Relat Res 2007; 463:237–43. doi: 10.1097/BLO.0b013e3180463089.
8. Banks DB, Hay SM. Intra-articular synovial haemangioma of the elbow in an adolescent male. Shoulder Elbow 2014; 6:57–9. doi: 10.1111/sae.12034.
9. Busa R, Adani R, Marcuzzi A, Caroli A. Acute posterior interosseous nerve palsy caused by a synovial haemangioma of the elbow joint. J Hand Surg Br 1995; 20:652–4. doi: 10.1016/S0266-7681(05)80128-7.
10. Akgün I, Kesmezacar H, Oğüt T, Dervişoğlu S. Intra-articular hemangioma of the knee. Arthroscopy 2003; 19:E17. doi: 10.1053/jars.2003.50068.
11. De Filippo M, Rovani C, Sudberry JJ, Rossi F, Pogliacomi F, Zompatori M. Magnetic resonance imaging comparison of intra-articular cavernous synovial hemangioma and cystic synovial hyperplasia of the knee. Acta Radiol 2006; 47:581–4. doi: 10.1080/02841850600767724.
12. Llauger J, Monill JM, Palmer J, Clotet M. Synovial hemangioma of the knee: MRI findings in two cases. Skeletal Radiol 1995; 24:579–81. doi: 10.1007/BF00204855.
13. Kim SJ, Cho SH, Ko DH. Arthroscopic excision of synovial hemangioma of the hip joint. J Orthop Sci 2008; 13:387–9. doi: 10.1007/s00776-007-1232-0.
14. Price NJ, Cundy PJ. Synovial hemangioma of the knee. J Pediatr Orthop 1997; 17:74–7. doi: 10.1097/01241398-199701000-00017.