Myopericytoma-An Alternate Cause of Persistent Knee Pain in Rheumatoid Arthritis

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Abstract

Rheumatoid Arthritis can present with consistent pain over peripheral joints. The manner of presentation of a subcutaneous tumour such as Myopericytoma may be very similar to that of an inflamed joint leading to the high frequency of it being overlooked and inadequately treated. Knowing the radiological and pathological differences will direct us in the right road to timely and adequate treatment.

Introduction

Myopericytoma (MPC), originally described as Hemangiopericytoma (HPC) in 1942,[1] was a term used to describe a subcutaneous tumour mainly seen in distal extremities. Previous reports have talked about the misdiagnosis of these tumours frequently as Sarcomas. In our particular field of interest, their manner of presentation which can closely resemble an inflamed joint, may be overlooked, hence leading to inadequate treatment of the tumour.

We present the case of a lady known to have Rheumatoid Arthritis with persistent pain at the knee joint. The differential diagnosis of Myopericytoma particularly in patient's with Inflammatory Arthritis will be described in this report.

Case

A 55 year old lady diagnosed with Rheumatoid Arthritis on treatment with Disease Modifying Antirheumatic Drugs (DMARDs) presented with consistent pain over the right knee for 2 years and swelling for 1 year. All the other joints were not swollen or tender. Local inspection of the right knee revealed a 1.5 cm diffuse swelling on the medial aspect of the knee joint with tenderness on palpation. An X-ray of the knee joint (Figure 1 A, B) showed medial tibiofemoral joint space narrowing, mild diffuse osteopenia and prominent cortices.

In view of persistent single joint symptom and normal inflammatory markers, an ultrasound and MRI were done. Ultrasound of the right knee (Figure 2 A, B) revealed a hypoechoic lesion with prominent vascularity in the deep soft tissue. MRI (Figure 3 A-C) showed a 10x7 mm ovoid lesion with internal vascularity adjacent to the medial margin of the patella.

With a clinical and radiological suspicion of a Glomus tumour, local excision and histopathology was done. This showed monomorphic oval to spindle shaped cells with scant cytoplasm. There were inconspicuous nucleoli, fine chromatin and multilayered concentric growth around blood vessels (Figure 4 A, B). There were also many thin walled vascular channels with hemangiopericytoma like pattern at the periphery.

Immunohistochemistry showed perivascular myoepithelial cells positive for smooth muscle actin (SMA) (Figure 5). A few cells were positive for S100. Blood vessels were positive for CD31 and CD34.

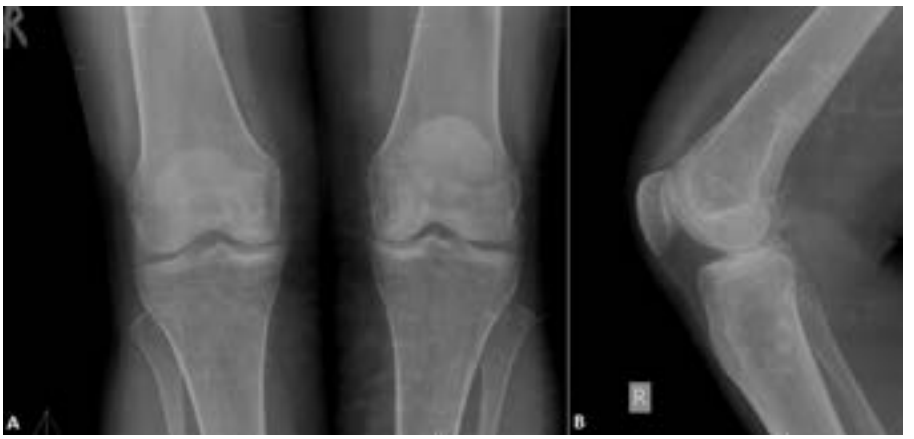

Fig. 1: (A) Anteroposterior and (B) right lateral radiographs of knee show mild diffuse osteopenia and prominent cortices. Mild medial tibiofemoral joint space narrowing with no obvious articular surface erosion or sclerosis. No suprapatellar bursal opacity on lateral view

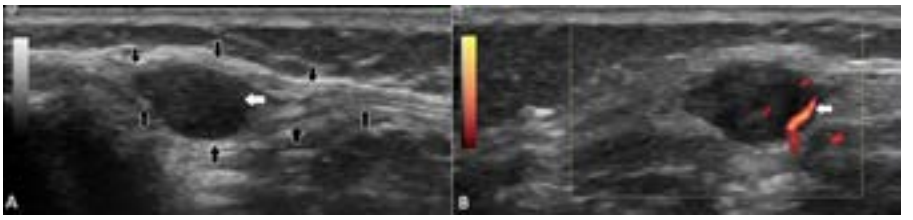

Fig. 2: (A, B) Longitudinal sonograms of right knee show a well defined oval homogenously hypoechoic lesion with eccentric prominent vascularity (white arrows) in the deep soft tissue of medial aspect of anterior knee. The lesion is seen within and splitting the medial retinaculum (black arrows)

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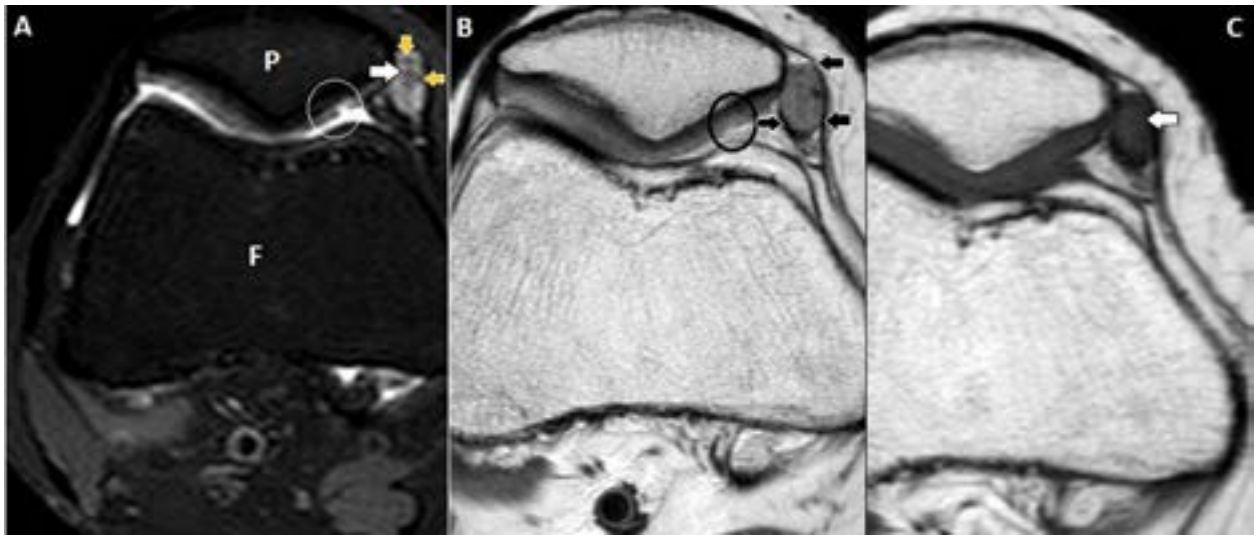

Fig. 3: (A) PD (proton density) fat suppressed axial, (B) PD-weighted axial and (C) T1-weighted axial MR images confirm the PD hyper and T1 hypointense lesion (white arrows) in the medial deep soft tissue aspect adjacent to patella splitting the fibres of medial patellar retinaculum and medial patellofemoral ligament (black arrows). Prominent hypointense dots (yellow arrows) within the lesion suggest intra-lesional vascularity. Note the full thickness defect in the mid part of medial patellar cartilage (circles). P-patella, F-femur

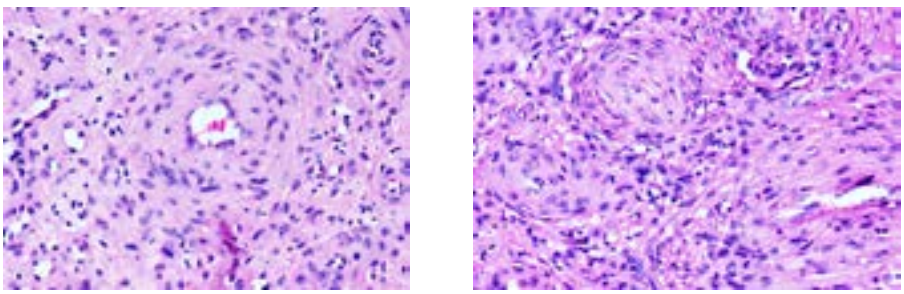

Fig. 4: (A, B) Haematoxylin and eosin stained; original magnification, 400x; Oval to spindle shaped cells with a multilayered concentric growth around blood vessels

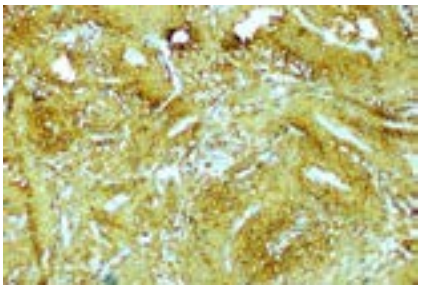

Fig. 5: Smooth muscle actin IHC, original magnification, 200x; The perivascular myoepithelial cells are positive for smooth muscle actin

Following local excision, she did not have recurrence of pain or swelling over the site. She continued DMARDs and her Rheumatoid arthritis is in remission.

Discussion

Myopericytoma is a benign solitary tumor most commonly seen in the dermis and subcutaneous soft tissue of the lower extremities. It is usually painless. It is primarily seen in middle aged adults and has a slightly more male predisposition. Rarely, it arises in the deep soft tissue of limbs. When it is located in the deep soft tissue of an extremity it can mimic other soft tissue masses like neurogenic tumor or glomus tumor or rarely other soft tissue sarcomas when it is large.[2] It is composed of oval to spindle shaped myoid appearing cells with a tendency for concentric perivascular growth.[3] It has overlapping features with other tumors such as glomus tumor, HPC, myofibroma and angioleiomyoma.[4] The rarity of this disease may be attributed to the possibility of under diagnosis. Prominent vascularity on MRI and colour Doppler ultrasound may help to differentiate myopericytoma from neurogenic tumour by revealing the hypervascular nature of the tumor. However, differentiation from glomus tumor preoperatively is difficult as both these lesions are hypervascular.

Table 1: (5)SMA-Smooth Muscle Actin; HPC-Hemangiopericytoma

Macroscopic features	Microscopic features
• It usually presents as a nodular, well circumscribed mass • Rarely exceeds 2cm in diameter	• Spindle and ovoid cells with Eosinophilic cytoplasm and elongated nucleoli • Characteristic multilayered growth of tumor cells around thin walled vessels • Surrounding vessels may present an extensive HPC like vasculature with large, irregular "staghorn" vessels
Immunohistochemistry	
• Positive • SMA*, h-caldesmon, CD 34 (focal)	• Negative • CD 31, S100 protein, Desmin, Cytokeratin

Histopathological features and immunophenotype are mentioned in Table 1.

It is treated by a simple surgical excision and has a low recurrence rate. Recurrence may occur due to poor circumscription of the lesion. Malignancies are rarely seen. It is important to note that in patients with inflammatory arthritis, a tumor can easily be overlooked. It is therefore important to be aware of the manner of presentation of such tumors in order to manage them adequately. This joint symptom can otherwise be managed as active joint inflammation, with unnecessary hike in immunosuppression and anti-inflammatory medications.

Conclusion

Diffuse swelling of a joint can be due to various causes one of which is a tumour such as Myopericytoma. Awareness of this entity is important as it can change a patients' plan of management.

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