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Case Report

Roca disease: An osteochondrosis of the inferior pole of the scapula with review of the literature ☆,☆☆

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ABSTRACT

Osteochondrosis is a developmental condition affecting the endochondral ossification. It is commonly idiopathic however can be due to vascular anomalies, dietary conditions, hormonal irregularity, or overuse trauma. Osteochondrosis occurring in the inferior pole of the scapula is an extremely rare condition and is referred to as roca disease. A higher degree of suspicion is required especially in a young patient with atraumatic shoulder pain and additional unconventional MRI sequences focusing on the inferior pole of scapula can be taken to rule out such conditions. We report a case of Roca disease in a 16-year male who presented with right shoulder pain. This is the third case report of roca disease in the English literature according to our knowledge and the first case report to demonstrate extensive MRI imaging features with a normal radiograph. Imaging, particularly MRI, plays a pivotal role in the diagnosis of this extremely rare entity. Also, the inferior pole of the scapula is usually not included in routine shoulder MRI imaging thus close scrutiny with additional MRI sequences should be done to diagnose such a rare entity.

Clinical history

A 16-year male came to the outpatient urgent care center with acute right shoulder pain and pain on the back between the shoulder blades with a decreased range of motion for 1.5 weeks. The pain was getting worse over the past 4 days. No history of trauma, bone disease, previous surgery, or fever. No history of relevant athletic or repetitive activities of the shoulders. On examination, vital signs were unremarkable; the local physical exam revealed no skin bruise, bleeding, swelling

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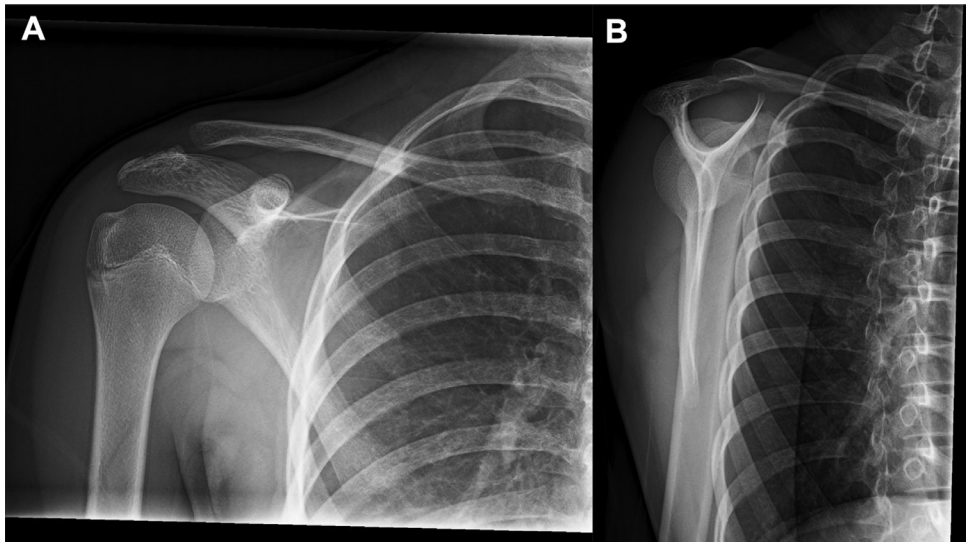

Fig. 1 – Right shoulder X ray in AP and (scapular Y) lateral views shows no abnormality.

seen along the right shoulder, or back of the torso. The overhead range of motion of the right arm was limited due to pain. Mild focal tenderness was elicited on the inferior angle of the right scapula. Routine labs including complete blood count, ESR, and CRP were unremarkable. The radiograph of the right shoulder was unremarkable (Fig. 1). MRI of the right shoulder was performed to rule out rotator cuff tear.

During MRI shoulder routine sequence acquisition, edema was partially visualized along the inferolateral right scapula and adjacent soft tissue in axial PD (proton density) and coronal T2 fat suppressed images (Fig. 2). Supplementary axial and coronal PD fat-suppressed images were obtained through the scapula, which showed edema centered at the secondary ossification center at the inferior pole of the scapula (Figs. 3 and 4). The patient was diagnosed with osteochondrosis and was treated conservatively with analgesics and activity restriction. The patient's pain decreased, and the range of motion improved in 2 months. Follow-up MRI was not obtained for comparison.

Table 1 – Osteochondrosis with eponyms.

Eponyms	Locations of osteochondrosis
Roca disease	Scapula lower pole
Panner disease	Humerus capitellum
Kienbock's disease	Lunate
Dieterich disease	Metacarpal head
Legg–Calvé–Perthes disease	Femoral head epiphysis
Osgood–Schlatter disease	Tibial tubercle
Sinding–Larsen–Johansson disease	Patella lower pole
Sever disease	Calcaneal apophysis
Köhler disease	Navicular bone
Islene disease	5th metatarsal base
Freiberg disease	2nd or 3rd metatarsal head
Scheurmann's disease	Vertebral body ring epiphysis

Discussion

Osteochondrosis is a developmental condition affecting the endochondral ossification in an immature skeleton [1,2]. Some of the common locations of the growing skeleton are listed on Table 1 [4,5]. Osteochondrosis of the inferior pole of the scapula is an extremely rare condition, known as roca disease [1,3]. Roca was the first to describe this condition in a 19-year-old amateur female basketball player and it was later also reported in a 14-year-old male amateur swimmer by Skaf and Taneja [3,6]. The pathogenesis of osteochondrosis is still unclear however vascular failure is believed to be the most likely cause. The theorized etiology for osteochondroses in various parts of the body include vascular failure due to vascular abnormalities, dietary factors, hormonal imbalances, anatomical abnormalities and repetitive microtrauma (perhaps due to overuse), which may also play a role in roca disease [7,8].

The ossification of the scapula occurs from seven centers of ossification by the age of 25 years. One of the ossification centers is located in the inferior pole of the scapula [3]. Skaf and Taneja mentioned that osteochondrosis in the roca disease occurs during the development of this ossification center between 14 and 25 years of age [3]. The active involvement in sports during this age may place undue stress on the inferior pole of the scapula resulting in overuse microtrauma [4]. Further, recent studies on animals have shown that insufficient vascular supply to the cartilage canal results in disruption of endochondral ossification [1,9].

As in our patient, the presentation of osteochondrosis is typically nonspecific, such as pain and decreased range of motion, requiring the physician to rule out trauma, infection, and inflammation [3,10]. Our patient complained of nontraumatic acute right shoulder pain, and pain between the shoulder blades, along with a decreased range of motion of the right shoulder for one and a half weeks. This is

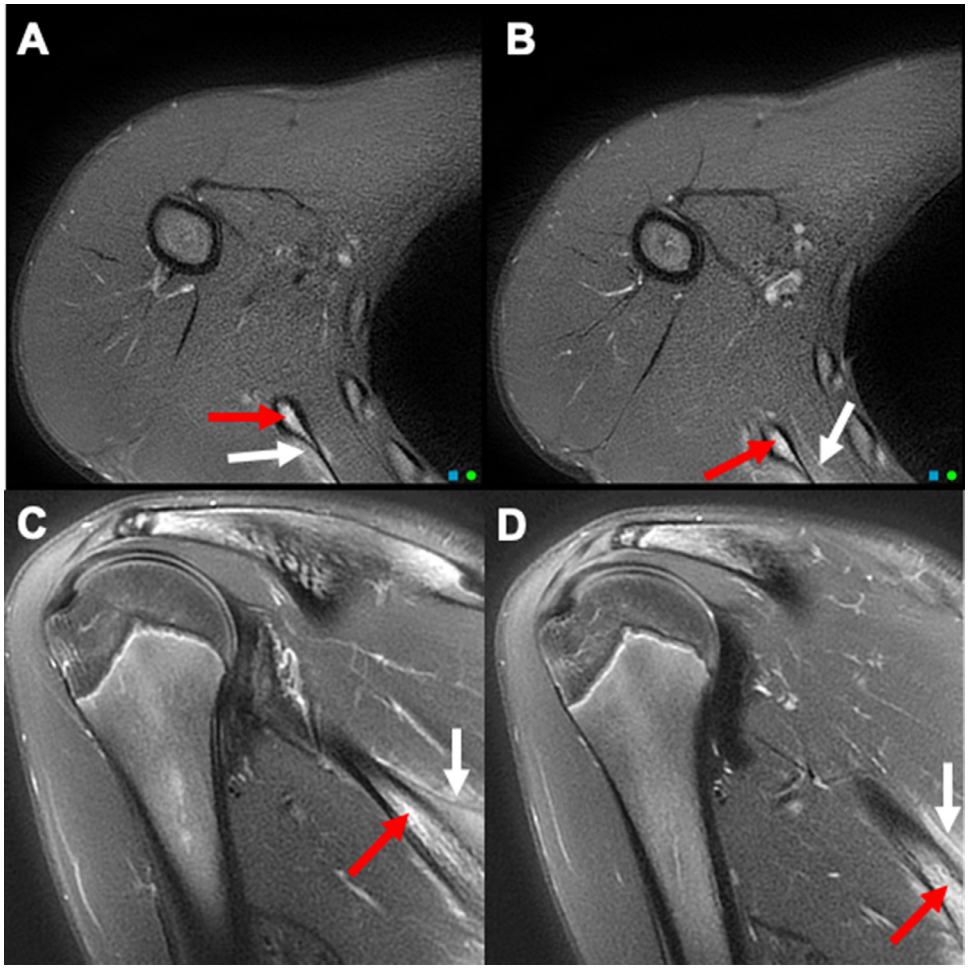

Fig. 2 – Right shoulder routine MRI axial PD (proton density) sequence (A and B) and coronal T2 fat suppressed sequence (C and D) shows partially visualized hyperintensity/edema involving the lower body of the scapula (red arrows) and adjacent soft tissue (white arrows).

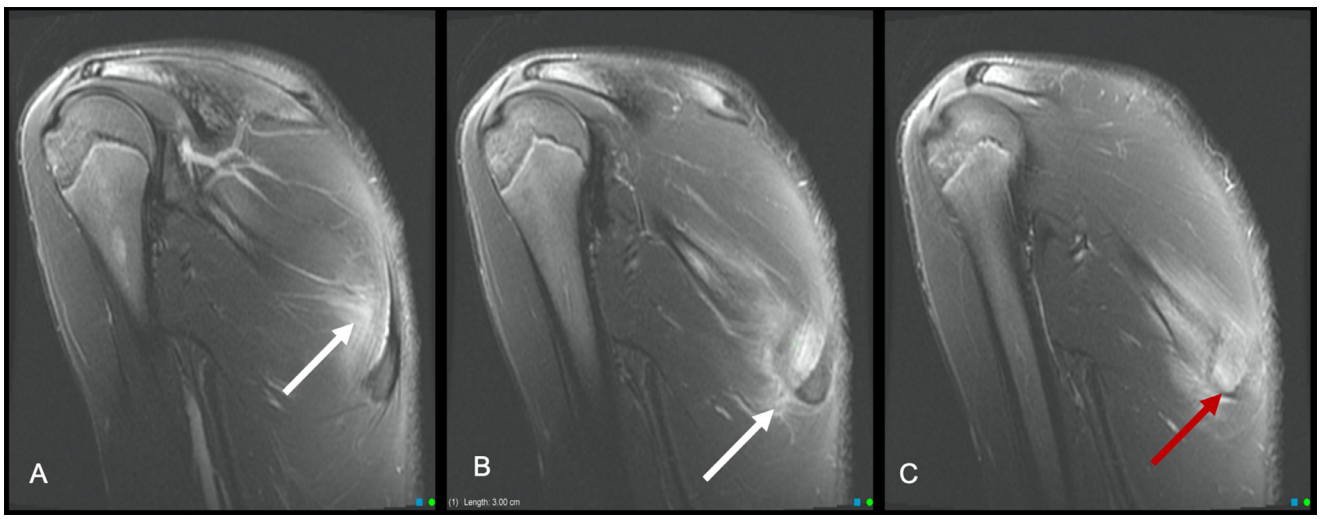

Fig. 3 – Oblique sagittal (proton density fat saturated) PD FS images of the right shoulder joint. Images A and B demonstrate soft tissue edema centered around the inferior angle of scapula (white arrows). Image C demonstrates edema involving the nonossified cartilaginous secondary ossification center at the inferior angle of scapula (red arrow).

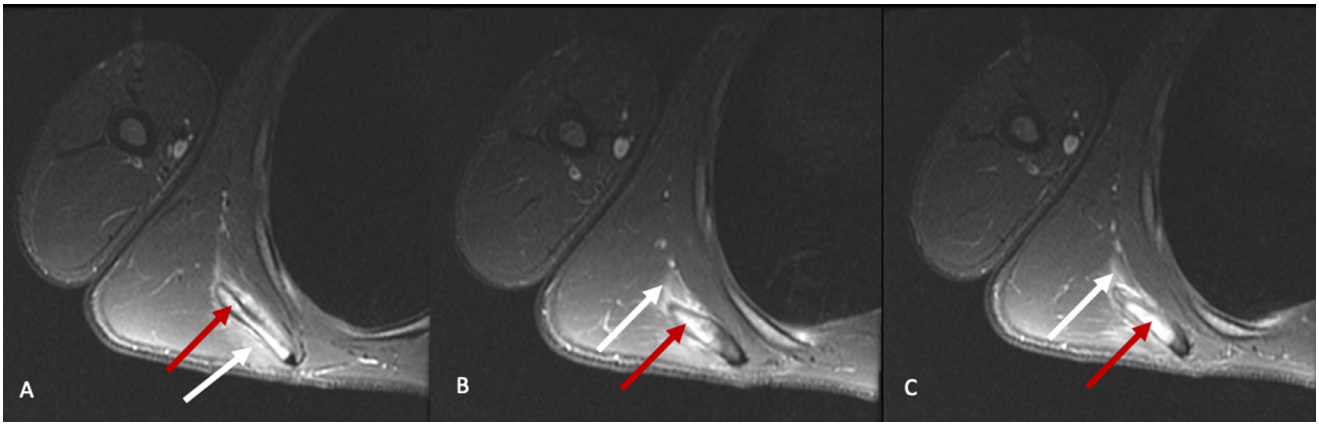

Fig. 4 – Axial (proton density fat saturated) PD FS images of the right shoulder joint. Images A, B, and C demonstrate edema involving the body of scapula (A and B) and nonossified cartilaginous secondary ossification center (C) at the inferior angle of scapula (red arrows) and the surrounding soft tissues (white arrows).

similar to the previously reported cases of roca disease and in accordance with the presentation of osteochondrosis [3].

Imaging findings reflect the various stages in the disease process, including necrosis, invasion of granulation tissue, revascularization, in the form of edema, inflammation, deformity, decreased perfusion, signs of repair, and revascularization [11]. Bilateral imaging is essential for a comparison of the affected side with respect to the normal side [11]. A radiograph may show irregularity, fragmentation, and sclerosis of the inferior angle of the scapula. However, CT scan and MRI are much helpful for better evaluation as a radiograph may be unremarkable in subtle cases [3,4]. CT scan helps delineate anatomy while MRI is sensitive for marrow edema and soft tissue abnormalities [3]. CT scan demonstrates sclerosis and heterogeneity in the ossification center and it also shows the decreased size of the affected scapula [3]. Volume rendering technique can demonstrate decreased size and irregularity of the affected bone, in comparison with the normal bone [3,4]. MRI is the most useful imaging modality to evaluate bone marrow, periosteal, and adjacent soft tissue edema. On MRI, the affected area appears low signal on T1 weighted images, bright on T2 weighted images, and shows postcontrast enhancement [3]. However, contrast enhancement might be absent if the affected segment is ischemic/necrotic [12]. The lower pole of the scapula usually not included in the field of view for the MRI scan, thus it is important to consider the possibility of roca disease and the field of view can be extended to include the lower pole of the scapula or additional sequences focused on the lower pole of the scapula like axial and coronal PD fat suppressed and/or STIR (Short tau inversion recovery) sequences should be obtained. Further, Technetium-99 bone scans would demonstrate increased uptake in the bone affected by osteochondrosis [4]. Resolution of the disease process is reflected in subsequent imaging studies in other locations of osteochondrosis, and we believe the same for the roca disease given the similar etiological factors but due to the limited literature on this rare entity it's still inconclusive [11,13,14]. If necrosis is not developed, radiograph shows complete recovery with no cortical abnormality, bone remodeling or bone loss and CT scan or MRI shows no cortical or signal abnormality. In case of developed necrosis in the affected endochondral ossification, sclerosis and fragmentations are seen in the CT scan and MRI however timely resumed vascularization likely resolve the condition in most of the cases [15].

One case of roca disease was successfully managed with conservative measures such as analgesics and restriction of activity with gradual re-introduction, while the other case required surgical management by excision due to failure of the initial conservative approach [3]. In the latter case, histopathological analysis of the excised bone tissue specimen demonstrated necrosis and fibrosis [3]. In case of conservative management, periodic follow-up is essential to evaluate the response [4]. Imaging may be used in follow-up to demonstrate the resolution of osteochondrosis [13].

Conclusion

It is essential for the physicians to be aware of Roca disease as a rare yet possible diagnosis in the patients presenting with shoulder pain or restriction of movement with pain around the shoulder blade region. A conventional radiograph may or maybe not sensitive enough to show the subtle changes, thus may necessitate investigating further with CT scan and MRI. Initial conservative management may be helpful; if conservative management fails, then surgery may be indicated [1,4]. Periodic follow-up is important in assessing the response of the patient and in determining the need for the change in approach [4].

Ethics approval and consent for publication

Not applicable.

Consent for publication

Not applicable.

Availability of data and material

Not applicable.

Authors' contributions

DG conceived the idea of this article and contributed to writing the manuscript substantially. KA contributed to the discussion of the manuscript substantially. HS contributed to figure formation of the manuscript and its revision. SS contributed to figure and manuscript revision. NG contributed to reference numbering and revision of the manuscript. RK contributed to design and revision of the manuscript.

Supplementary material

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2020.06.042.

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