Case report

Bilateral trochlear avascular necrosis: A case report and brief review of the literature

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ABSTRACT

Introduction and importance: Trochlear AVN is a very rare entity with only a limited number of cases being reported.

Case presentation: Our case is interesting as it presents a case of an incidentally found bilateral trochlear AVN in a 12 year-old child when he presented with an undisplaced right supracondylar fracture.

Clinical discussion: It is highly likely that an unknown underlying predisposing factor, possibly recurrent microtrauma, was responsible for the bilateral occurrence.

Conclusion: We recommend further research to identify other etiological and predisposing factors, as the etiological factors still remain unclear, despite the available literature.

1. Introduction

Hegemann’s disease, or avascular necrosis (AVN) of the trochlea is a rare complication after fractures about the elbow, usually following a supracondylar, a lateral condyle, a medial condyle, or after Salter–Harris type I epiphyseal fractures in childhood [1,2]. These terms are described to be a spectrum of poorly defined vascular disorders of the distal humerus, varying from a benign mild vascular disorder to a complete avascular necrosis after fractures, and from a fishtail deformity to a dissolution of the humeral trochlea [1,2].

Debate still exists in regard to the etiology as well as the management of trochlear AVN. Our current case describes a case of idiopathic bilateral trochlear AVN that was incidentally found when the child presented with an undisplaced supracondylar fracture after a trauma to his right elbow.

This case report has been reported in line with the SCARE 2020 criteria [3].

2. Case

An overweight (BMI of 27) previously healthy medically and surgically, not on any medication, 11 year-old boy presented to the emergency department at King Saud University Medical City in Riyadh (a tertiary healthcare facility) with a history of a trauma to the right elbow 2 h prior to his presentation. His friend fell on his outstretched right arm. Patient was seen and assessed by the orthopedic on-call team. He was vitally stable and had no previous traumatic history, motion limitation, pain, locking, nor prior infection in either elbow. Local examination of his elbow revealed a right elbow swelling without deformity or instability. Tenderness was noted over the lateral condyle. Range of motion (ROM) was 0–90 of elbow flexion-extension and he was able to do full pronation and supination. Distal neurovascular status was intact. His left elbow exam was negative for swelling, tenderness, deformity, as well as instability. He had a full left elbow ROM. His initial right elbow X-ray (Image 1) showed a posterior fat pad sign, with no clear fracture line, but an abnormally appearing trochlea was evident. Afterwards, X-rays of the contralateral elbow (Image 2) showed a similar abnormal appearance of the trochlea. Carrying angles were 9 and 5° in valgus, for right and left elbows, respectively. The trochlear notch angles were 123 and 112°, for right and left elbows, respectively. He had no complaints from his left elbow, therefore, he was discharged on an above elbow cast for the right arm with a close follow-up and analgesia. The parents were in agreement with our assessment and treatment plan. During his follow-up, he was found to have an undisplaced supracondylar fracture in addition to the diagnosis of bilateral Hegemann’s disease (Image 3). A repeat history was found to be negative for preceding trauma. After cast removal (at 6
Image 1. AP and lateral radiographs of the right upper limb at the time of the trauma.

Image 2. Radiographs of the left upper limb at the time of the trauma.

Image 3. Radiographs of the right upper limb 2 weeks after the trauma.
weeks) (Image 4), ROM exercises at home were advised to improve his range of motion. Following the readings of his carrying angles and trochlear notch angles was not possible due to the inappropriate X-ray views that were done in clinic. He is scheduled for new X-rays in his next clinic visit.

During his latest follow up (12-week visit), his right elbow showed no tenderness, and ROM was 30–110, and was referred for physical therapy to improve his ROM. During all his visits, his left elbow revealed no deformity and he had a full painless ROM. The patient is still being seen in our clinic for motion arc assessments as well as possible improvement of his bilateral trochlear avascular necrosis (AVN).

3. Discussion

Avascular necrosis is a condition that leads to cellular death of the bone as a result of temporary or permanent interruption of blood supply. Its occurrence around the elbow is relatively uncommon, and even less common to occur in the trochlea, compared to the capitellum, radial head, and olecranon [4]. The reported incidence of Hegemann's disease is 0.001–0.27% [4,5].

Definitive causes behind the development of Hegemann's disease have not been identified [6]. However, several traumatic and atraumatic associations have been definitely linked to trochlear osteonecrosis including acute or remote trauma, such as fractures, chronic repetitive microtrauma, and contusions, the administration of chemotherapeutic agents, as well as being idiopathic in nature [1,2,4,].

The possible differential diagnosis of elbow pain in children and younger patients should include osteochondritis dissecans (found more frequently in boys compared to girls and more on the right side compared to the left and from the clinical viewpoint, it commonly reveals joint locking, as opposed to trochlear AVN), tuberculosis (especially in the 2nd or 3rd decades of life), fungal infections (mainly presents with olecranon bone atrophy on imaging, and shows no reactive changes), syphilis or lues (involves both elbows in children), osteomyelitis, tumors, osteonecrosis (may involve the olecranon, epicondyles, trochlea, humeral capitulum, head of the radius), epiphysiodesis, septic and rheumatoid arthritis, osteoarthritis (most post-traumatic when seen in children), chondromalacia, bursitis, radial tunnel syndrome, annular ligament syndrome, ulnar neuritis, triceps tendinitis, instability and epicondylitis and tendonitis of the extensor musculature [5,7].

Since our patient did not have joint locking, present with fever or pain with passive ROM, present with history of previous trauma or fractures, and the presence of trochlear changes on X-ray, we were able to deduce that our patient most likely had trochlear avascular necrosis, possibly due to recurrent microtrauma, as he constantly wrestles.

The trochlear epiphysis ossification center appears after the age of 5 years, developed between the ages of 8 and 13 years in boys, and fuses with the humeral metaphysis between the age of 13 and 16 years [1,2]. The pediatric elbow is said to be supplied by a complicated anastomotic network of vessels. An extraosseous and an intraosseous system supplies the area around the distal humerus [2]. Therefore, the general consensus that was found is that osteonecrosis of the trochlea involves collapse of both bone and articular cartilage due to the lack of blood supply [2].

Trochlear AVN has been described to occur either partially or completely. Type A (partial involvement), has been described to affect the apex or the lateral portion of the trochlear medial crista. Those patients are usually asymptomatic and present without any angular deformities. Their characteristic radiologic findings are a central deficiency of the distal humeral epiphysis. Whereas Type B (complete involvement), has been described to involve the whole trochlear metaphysis. Those patients tend to present with progressively developing elbow varus deformity and significant loss of ROM [4,8,9]. Hegemann’s disease is specifically said to undergo five stages of development on radiographs based on Schumacher et al. [10]. Stage 1 shows initial loss of density and later plaque-shaped sclerosis of the centre of epiphysical ossification; stage 2 shows reduction in size and condensation of the ossification centre; stage 3 shows loosening, accompanied by onset of new ossification; stage 4 shows regeneration and enlargement of the ossification centre; and stage 5 shows final stage (complete or partial recovery) [5,9]. Our current case can be classified as a type A trochlear AVN, due to the subtle and partial involvement of the trochlea.

Beyer et al. demonstrated that aseptic necrosis of the trochlea showed a low intensity signal on T1-weighted MRI images, and reported the usefulness of MRI in diagnosing Hegemann’s disease as well as in verification of recovery [5]. In our patient, MRI was not done due to the COVID-19 situation in our hospital, where only urgent cases are currently being accepted and scheduled.

Uhrmacher et al. [11] was the first to describe Hegemann’s disease in 2 children aged 7 and 9 years. The main presenting symptoms were swelling and restricted elbow ROM. Which is similar to that of other studies that ranged from pain [6,7,12], crepitus [12] and restricted ROM [1,12].

Ott et al. [12] brought up the spectrum of operative intervention that may be offered to a patient, and that depended on the individual pathology, and ranged from arthroscopic or open debridement with capsulotomy, surgical arrest of the remaining medial or lateral epiphysis, to osteotomy for persistent deformity [12]. Claessen et al. [1] recommended conservative treatment in most cases, in the form of rest, less physical activity, or avoidance of vigorous sport activity. Pain resolution was documented in most children with Hegemann’s disease. In contrast,86% of those found to have a fishtail deformity had pain and limited ROM. In patients who underwent operative treatment, pain relief was seen in 85% and improvement in ROM was seen in 100% of the
involved patients [1]. Hara et al. and Alici et al. report conservative management to be of benefit [6,7]. Bayer et al. reports varus deformity and limited range of motion and subsequently, trochlear AVN to be one of the sequelae of conservative management [5] which is in agreement with other papers [2,4]. In our case, potential expected complications include elbow stiffness and deformity. For the stiffness, we initially started home exercises and when no improvement was noted, we asked for a referral to physical therapy to improve his arc of motion. For such, we would suggest continued ROM observation. In regards to the deformity, we would suggest observation with serial radiographs until full growth potential is achieved.

In contrast to the available literature, our case presents a single patient with incidentally found bilateral trochlear AVN that was painless in nature, except for the superimposed sustained undisplaced supracondylar fracture, and did not show limitation of movement as evidenced by the normal ROM on the unaffected upper limb. In our case, ROM was initially $0^\circ–90^\circ$, and after cast removal was $30^\circ–110^\circ$. The decrease in his right elbow extension can possibly be due to elbow stiffness development due to immobilization. For such, the patient is currently undergoing physical therapy for his elbow to improve that ROM.

There are several areas of weakness in our study. First, it is a case report, which is not the strongest form of evidence. Second, it is important to note that in the pediatric population, traumatic events may sometimes be overlooked, missed, or even under-reported. Our patient may have had prior several microtraumas to both elbows that may have been passed as contusions or other forms of minor trauma. In addition, we are still following this patient, and as such, development of progressive varus is still not evident and cannot be definitely stated until his growth potential has been fully achieved. Based on our findings, we recommend having a low threshold to obtain contralateral elbow radiographs in the case where a fishtail deformity or trochlear AVN is noted and to use for comparison, as the appearance of the growth plate of the trochlea may differ between individuals. Moreover, we recommend further research to identify other etiological and predisposing factors. In addition, we recommend studying the possible long-term effect of arthritic debridement in possibly preventing the future occurrence of osteoarthritis associated with Hegemann’s disease and fishtail deformities of the elbow.

4. Conclusion

Trochlear AVN is a very rare entity with only a limited number of cases being reported. Our case is interesting as it presents a case of incidentally found bilateral trochlear AVN. It is highly likely that an unknown underlying predisposing factor was responsible for the occurrence of the bilateral finding, as our patient denied a history of trauma to the left elbow, and only presented with a right type 1 supracondylar fracture. We highlight the possibility of repetitive microtrauma occurring with contact sports such as wrestling and others, which may predispose to trochlear AVN. We recommend further research to identify other etiological and predisposing factors, as the etiological factors still remain unclear.

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Author contribution

Khalid Bakarman: Revising the article critically for important intellectual content and final approval of the version to be submitted.

Nayef Alghnimei: Revising the article critically for important intellectual content and final approval of the version to be submitted.

Sultana Borai: The study concept, design, acquisition of data, interpretation of data, drafting the article and revising it critically for important intellectual content

Rheema Alfadhil: The study concept, design, acquisition of data, interpretation of data, drafting the article and revising it critically for important intellectual content

Research registration

Not required, as our article does not describe a new surgical technique or new equipment/technology.

Patient informed consent

Written informed consent was obtained from the patient’s legal guardians (parents) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Khalid Bakarman, Nayef Alghnimi, Sultana Borai, Rheema Alfadhil.

Declaration of competing interest

The authors in this case study report no potential conflict of interest relevant to this article.

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