

Arthroplasty in Ochronosis “Tips and Pearls in Surgery”: Case Series

Cagatay Ulucay^{1*}, Turhan Ozler¹, Faik Altintas¹, Muharrem Inan² and Abdurrahman Onur Kocadal¹

¹Yeditepe Medical Faculty, Orthopaedics and Traumatology Department, Ankara Cad no 102 Kozyatagi Istanbul, Turkey

²Cerrahpasa Medical Faculty, Orthopaedics and Traumatology Department, Cerrahpasa No 1 Fatih Istanbul, Turkey

Abstract

Introduction: The aim of this paper is to get the surgeon familiar with black cartilage and massive synovitis while performing a total joint replacement operation including preoperative, intraoperative and postoperative tips and pearls.

Case presentation: Two Caucasian male patients, one with knee osteoarthritis and one with both knee and hip osteoarthritis.

Conclusion: The black cartilage and softened bone does not heal different from the normal bone, therefore internal fixation of the fractures and cement less fixation of the prosthesis will not fail. Caution should be paid for excessive bleeding after total synovectomy. Patients have to be informed about consecutive operations and recurrent postoperative effusions.

Introduction

Ochronosis is a known disease since Virchow figured out the clinical presentation in 1866. Reid published the cartilage disorder involving with early onset osteoarthritis [1]. Alkaptonuria is characterized by deposition of polymerized homogentisic acid pigment in connective tissue and is called as ochronosis [2]. Ochronosis arises in approximately 50% of alkaptonuric patients [3]. The lack of homogentisic acid oxidase enzyme is the reason of this deposition. It is a rare autosomal recessive metabolic disorder which has an incidence one per 2-5 million live births [4]. The clinical presentations of the disease are dark/black pigmentation of connective tissues, cartilage destruction with early onset osteoarthritis, recurrent effusions, degenerative spine with/without ne-dovo scoliosis, dyspnea, coronary and valvular calcifications and disorders, urinary calculi and scleral pigmentations [5,6]. There is no permanent cure for the disease but anti oxidant drugs and gene therapies are still investigating [7]. The treatment management is usually symptomatic and most of the patients refer to the clinics for orthopaedic problems.

The reason of publishing this paper is to inform orthopaedic surgeons about intraoperative pitfalls and unproven myths of ochronosis with our personal experiences and some review of the latest literature.

Case Presentation

Case 1

A 56-year old Caucasian male patient was admitted to our clinic with history of bilateral knee and low back pains for 5 years. On physical examination, there were gray-black pigmentation zones in his sclera and on his ear helices. Range of motion of the right knee was 0°–10°–90° and left knee was 0°–0°–110°. Lysholm scores were 33 for the right knee and 59 for the left (<68 Bad). Lumbar and thoracic vertebral physical examination showed restricted and painful range of motion. Qualitative urine tests (FeCl₃ and Benedict) showed high levels of HGA and thus confirmed diagnosis of ochronopathic arthropathy. Patient underwent a cemented right total knee arthroplasty (Genesis II, Smith & Nephew, Memphis). The procedure was performed under tourniquet control. It was our first time we all saw black cartilage after capsulotomy. Black colored femur and tibia joint cartilages and dark colored massive synovial hypertrophy were observed (Figure 1).

Standard cemented total knee arthroplasty with aggressive total synovectomy was performed. No complications occurred during the surgery. At the recovery room, the anesthesiology specialists warned us about the unstable hemodynamic condition of the patient. Within the first postoperative hour, a continuous 1500 cc bleeding into the drain was observed. It resulted with uncontrolled hypotension and tachycardia. Two units of erythrocyte suspensions immediately transfused with 2000 cc lactated ringer solution. His dressing was renewed because of the excessive bleeding from the wound. We decided to explore the knee for a major vascular damage if bleeding had been continuing. Pressured bandages and cold application was performed. The bleeding slowed down within 10 minutes while the patient hemodynamically stabilized.



Figure 1: Intraoperative appearance of gray-black colored femur and tibia cartilages and dark colored massive synovial hypertrophy.

***Corresponding author:** Cagatay Ulucay, Yeditepe Medical Faculty, Orthopaedics and Traumatology Department Devlet Yolu Ankara Cad no 102 Kozyatagi Istanbul, Turkey, Tel: +905327284719; E-mail: culucay@turk.net

Received November 11, 2013; **Accepted** December 03, 2013; **Published** December 11, 2013

Citation: Ulucay C, Ozler T, Altintas F, Inan M, Onur A, et al. (2013) Arthroplasty in Ochronosis “Tips and Pearls in Surgery”: Case Series. J Arthritis 3: 116. doi:10.4172/2167-7921.1000116

Copyright: © 2013 Ulucay C, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

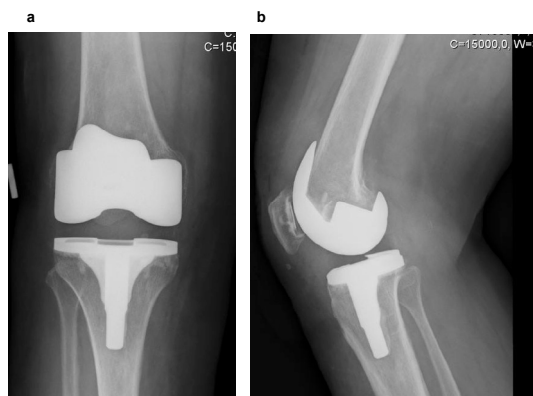


Figure 2: 3rd postoperative year X-ray.

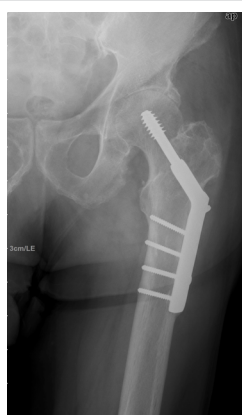


Figure 3: Healed proximal femoral fracture which was operated with internal fixation using sliding-screw plate.



Figure 4: Gray-black pigmentation zones on sclera and ear helices of the patient.

At the intensive care unit, 3 more units of erythrocyte suspensions were also transfused. The patient stayed at the intensive care unit for 1 day, which is an unusual condition for a total knee operation of a considerably young patient. At the third year follow-up visit there was no complain in right knee and Lysholm score was 88 (77-90 Good) (Figures 2a, 2b). The patient didn't want any operation for the left knee although there was a severe osteoarthritis. This may be the reason of his intensive care unit stay with a risk of exsanguination.

Case 2

A 60-year old Caucasian male patient was admitted to our outpatient clinic with left hip pain and restricted hip motion. Patient had a history of traffic accident 12 years ago and sustained a proximal femoral fracture, which was treated with sliding-screw plate (Figure 3).

8 years ago, patient was also admitted to another clinic for knee osteoarthritis. He had cemented total knee arthroplasty operations for his both knees. On physical examination, there were gray-black

pigmentation zones in his sclera and on his ear helices (Figure 4). Patient's Harris hip score was 26 (<70 Bad). Quantitative HGA level was 122-mmol/mol creatinin (normal: 0) in gas-liquid chromatography and thus confirmed diagnosis of ochronopathic arthropathy. The patient prepared for cement less total hip arthroplasty. In lateral decubitus position, first sliding-screw plate system was removed. Then with modified Gibson approach, hip arthroplasty operation was performed. Dark brown-black colored joint cartilage was observed on femur and acetabulum after dislocation of the hip (Figures 5a, 5b).

Stable acetabular and long full porous femoral stem were fixed cement less without any complications (Synergy, Smith & Nephew, Memphis). In the first postoperative day, full weight bearing was allowed. At the 3rd year postoperative follow-up visit patient was able to walk with full weight bearing without any complaints. His Harris Hip score was 94 (90-100 Excellent) while he had severe complaints in his right hip. His right Harris hip score was 48 (<70 Bad). He was operated for his right hip with cement less acetabular and proximal porous anatomical femoral stem (Anthology, Smith & Nephew, Memphis), without any complications. At the 2nd year postoperative follow-up visit, patient was walking with full weight bearing without any major complaints at his four operated joints apart from having seldom recurrent effusions in his both knees (Figure 6). His Harris Hip score was 94 (90-100 Excellent) although he had some complaints of low back pain.

Discussion

Ochronopathic arthropathy is a rare, developed in alkaptonuria patients as a result of deposition of polymerized homogentisic acid pigment in the cartilages and synovial tissue [7-9]. Life-table analyses show that, patients start to have joint replacement operations at a mean age of 55 years, which is comparable with our patients as they were 56 and 52 years old respectively [3]. The disease intensifies after the age of

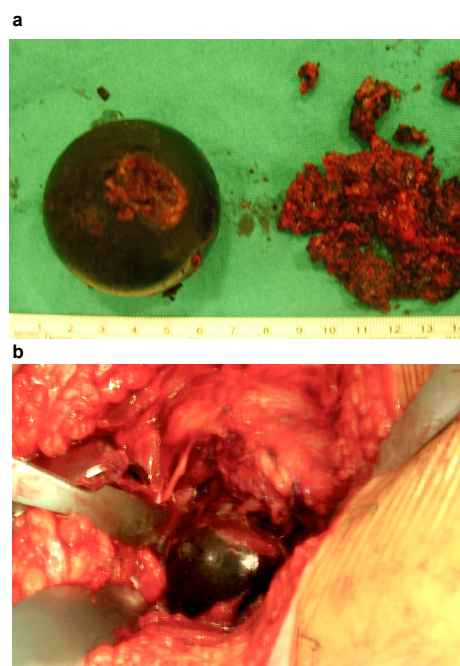


Figure 5: Intraoperative appearance of dark brown-black colored joint cartilage of femur and acetabulum.



Figure 6: Both hips and knees had operated.

30 years, with a more rapid increase in men than in women, which is also compatible with our patients as both are men [3].

Although bone tissue is partially affected by ochronotic pigmentation, in general bone formation and union is normal [10-13]. This is also proven with one of our cases; normal bone healing was observed following internal fixation of proximal femoral fracture with sliding-screw plate system. And also bone-in-growth is not affected and cement less prosthesis can be used in eligible cases [8,11,14]. We used cement less femoral stems and acetabular components in one of our cases and we didn't encounter any stabilization and early loosening problems. Both patients were able to weight bear on first postoperative day and there were no signs of early loosening, sinking or protrusion.

In one of our cases, we assumed that, massive postoperative bleeding was related to ochronosis. But our researches had shown that there are not any coagulation anomalies in ochronosis [7,15]. Our current experience suggests that, the reason of bleeding was aggressive total synovectomy, which was performed under tourniquet. Since it was our first experience with massive and dark colored synovial hypertrophy, we excised it all without any control. Total synovectomy should be avoided without intraoperative bleeding control.

Patients with ochronosis may have cardiac problems, which are coronary or valvular calcifications, and may have dyspnea and diminished vital capacity [16,17]. Therefore spinal anesthesia could be a good choice but degenerative changes such as narrowed disk spaces and spine fusions may challenge the regional anesthesia techniques [18]. For all our operations, anesthesiology specialists performed general anesthesia without any complications. Therefore we found general anesthesia also safe with proper preoperative preparations.

Conclusion

Although ochronotic arthropathy is a chronic disease with multiple joints involvement without any certain treatment, it is possible to relieve long lasting painless joints with properly performed joint reconstructions to improve patient's quality of life. The black cartilage and softened bone does not heal different from the normal bone, therefore internal fixation of the fractures and cement less fixation of the prosthesis will not fail. Caution should be paid for excessive

bleeding after total synovectomy. Patients have to be informed about consecutive operations and recurrent postoperative effusions.

Consent Section

Written informed consent was obtained from both patients for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References

1. Reid E: Ochronosis. *Proceedings of the Royal Society of Medicine* 1908, 1:57-58.
2. Drakoulakis E, Varvitsiotis D, Psarea G, Feroussis J (2012) Ochronotic arthropathy: diagnosis and management: a critical review. *Am J Orthop (Belle Mead NJ)* 41:80-83.
3. Phornphutkul C, Introne WJ, Perry MB, Bernardini I, Murphey MD, et al. (2002) Natural history of alkaptonuria. *N Engl J Med* 347:2111-2121.
4. Zacharia B, Chundarathil J, Ramakrishnan V, Krishnankutty RM, Veluthedath R, et al. (2009) Black hip, fracture neck of femur and scoliosis: A case of ochronosis. *J Inherit Metab Dis* 32 Suppl 1:S215-220.
5. Collins E, Hand R (2005) Alkaptonuric ochronosis: a case report. *AANA J* 73:41-46.
6. Keller JM, Macaulay W, Nercessian OA, Jaffe IA (2005) New developments in ochronosis: review of the literature. *Rheumatol Int* 25:81-85.
7. Abimbola O, Hall G, Zuckerman JD (2011) Degenerative arthritis of the knee secondary to ochronosis. *Bulletin of the NYU hospital for joint diseases* 69:331-334.
8. Aynaci O, Onder C, Turhan AU (2000) Bilateral hip arthroplasty for ochronotic arthropathy. *Clinical rheumatology* 19: 150-152.
9. Cetinus E, Cever I, Kural C, Erturk H, Akyildiz M (2005) Ochronotic arthritis: case reports and review of the literature. *Rheumatol Int* 25:465-468.
10. Konttinen YT, Hoikka V, Landtman M, Saari H, Santavirta S, et al. (1989) Ochronosis - a Report of a Case and a Review of Literature. *Clinical and experimental rheumatology* 7:435-444.
11. Aydogdu S, Cullu E, Ozsoy MH, Sur H (2000) Cementless total knee arthroplasty in ochronotic arthropathy: a case report with a 4-year follow-up. *J Arthroplasty* 15:539-543.
12. Skuginna A, Bockmann G (1988) [The ochronosis disease picture and conservative and operative therapy. Presented in relation to a case report]. *Z Orthop Ihre Grenzgeb* 126:643-646.
13. Aliberti G, Pulignano I, Schiappoli A, Minisola S, Romagnoli E (2003) Bone metabolism in ochronotic patients. *J Intern Med* 254:296-300.
14. Kerimoglu S, Onder C, Aynaci O, Malkoc CH (2005) Hip arthroplasty for ochronosis. *Saudi Med J* 26:1812-1814.
15. Kotela A, Pirkko K, Kotela I (2010) [Ochronosis as a cause of multiple joint osteoarthritis in one patient]. *Przegl Lek* 67:427-431.
16. Kragel AH, Lapa JA, Roberts WC (1990) Cardiovascular findings in alkaptonuric ochronosis. *Am Heart J* 120:1460-1463.
17. Wauthy P, Seghers V, Mathonet P, Deuvaert FE (2009) Cardiac ochronosis: not so benign. *Eur J Cardiothorac Surg* 35:732-733.
18. Ogata J, Tamura K, Miyamishi K, Minami K, Haranishi Y (2008) [Anesthesia in a patient with alkaptonuric ochronosis for total hip arthroplasty]. *Masui* 57:439-442.