



Total knee arthroplasty in ochronosis: a case report and critical review of the literature

Okronoziste total diz artroplastisi: Olgu sunumu ve kritik literatür incelemesi

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Alkaptonuria is an autosomal recessive disorder caused by the deficiency of homogentisate 1.2 dioxygenase activity. The clinical presentation shows an ochronotic pigment which is deposited in all connective tissues, including in cartilage, particularly. The knee is the most common site of peripheral abnormality. There is currently no definitive cure for alkaptonuric ochronosis. In this article, we present a 69-year-old male case who underwent bilateral cemented total knee arthroplasty simultaneously. Our results during two-year follow-up were satisfactory. A critical review of the literature revealed no uniformity in reporting such cases.

Key words: Alkaptonuric ochronosis; knee replacement arthroplasty; ochronosis.

Alkaptonüri, homogentisat 1.2 dioksijenaz enzimi aktivitesindeki eksikliğe bağlı gelişen otozomal resesif geçişli bir hastalıktır. Klinik tabloda kıkırdak başta olmak üzere tüm bağ dokusunda biriken okronotik pigment birikimi görülür. Periferik anormalliğin en sık görüldüğü yer diz eklemidir. Günümüzde alkaptonürik okronozisin kesin bir tedavisi yoktur. Bu yazıda, aynı seansta iki taraflı çimentolu total diz artroplastisi uygulanan 69 yaşında bir erkek olgu sunuldu. İki yıllık takip sonuçları tatmin ediciydi. Kritik literatür incelemesinde bu tür olgularda standart bir yaklaşıma rastlanmadı.

Anahtar sözcükler: Alkaptonürik okronozis; diz replasmanı artroplastisi; okronozis.

Alkaptonuria (AKU) is an autosomal recessive disorder caused by deficiency of homogentisate 1,2 dioxygenase (HGO) activity.^[1] In this disease ochronotic pigment is deposited in all connective tissues, but especially in cartilage. Simultaneously, this discoloration can occur in tendons, ligaments, sclera, heart valves, the intima of blood vessels, and the skin. The knee is the most common site of peripheral abnormality. Other sites of involvement are hips, shoulders, sacroiliac joints, and pubic symphysis.^[2]

There is currently no definitive cure for alkaptonuric ochronosis. However, total joint replacement in published cases of ochronotic osteoarthritis report good results similar to osteoarthritic patients without ochronosis. A careful review of the literature revealed only 21 knees in 13 cases (Table I). Because all these are case reports, no guideline is available for

replacement of the knee joint in ochronotic patients. Here we report a new case and attempt to perform a critical analysis of the literature.

CASE REPORT

A 69-year-old male presented with a 10-year history of pain in both knees. He weighed 75 kg, with a height of 155 cm. On physical examination, range of motion of the right knee was 0 to 110 degrees, left knee was 0 to 114 degrees.

Radiologic evaluation revealed characteristic findings of osteoarthritis, including narrowing of joint spaces, subchondral sclerosis, irregularities of the joint surfaces, and peripheral new bone formation (Figure 1). The patient was otherwise healthy.

The patient underwent bilateral cemented total knee arthroplasty. During surgery, dark discoloration

TABLE I

Comprehensive data of published literature

Article	Year*	Age/gender	PT	Side	FU	Pre ROM	Post ROM	Pp KS	Pp FS	Results*
Carrier and Harris ^[5]	1990	7/M	-	Bilateral	-	-	-	-	-	Benefit
Ramsperger et al. ^[18]	1994	57/F	-	Left	-	0-12	0-100	-	-	-
Konttinen et al. ^[7]	1989	58/M	C(-)	Bilateral	-	5-90	-	-	-	Good
Fisher and Davis ^[9]	2004	69/F	-	Bilateral	5 years	-	-	-	-	Improvement
Abimbola et al. ^[13]	2011	48/M	C(+)	Left	2 years	0-130	0-110	-	-	Excellent
Kotela et al. ^[6]	2008	59/M	C(+)	Bilateral	-	-	-	-	-	Good
Araki et al. ^[14]	2009	56/M	C(-)	Bilateral	-	-	-	-	-	Good
Aydogdu et al. ^[8]	2000	48/M	C(-)	Left	4 years	25-125	5-120	35-85	45-50	Good
Demir ^[4]	2003	70/M	C(+)	Bilateral	14 years	-	-	-	-	Good
Kefeli et al. ^[15]	2008	60/F	C(+)	Bilateral	10 months	0-100	0-100	-	-	Good
Fontao-Fernández et al. ^[16]	2010	68/F	-	Left	-	-	-	-	-	-
Spencer et al. ^[17]	2004	53/F	-	-	7 years	-	-	-	-	Good
Moslovac et al. ^[12]	2003	70/M	C(+)	Bilateral	7 years	-	-	-	-	Best

* Publication date; PT: Prosthesis type; FU Follow-up; Pre ROM: Preoperative range of motion; Post ROM: Postoperative range of motion; Pp KS: Pre- and postoperative knee score; Pp FS: Pre- and postoperative function score; + As stated by the authors; - Not reported; C(-): Cementless; C(+): Cemented.

was observed, with black gray discoloration on all cartilaginous surfaces and bone eburnations. The synovia was hypertrophic and black-brown in color. Histologic evaluation confirmed the diagnosis of ochronotic arthropathy.

After surgery, the patient was allowed weight-bearing as tolerated and physiotherapy was started on the first postoperative day. The patient underwent standardized inpatient physiotherapy during his hospital stay, including continuous passive motion, active-assistive and active range of motion exercises, isometric and isotonic strengthening exercises, gait training and transfer training, followed by a home-based exercise program. Hospital for Special Surgery (HSS) knee score and range of motion (ROM) of the patient were assessed preoperatively and at 6, 12, 26, 52 weeks postoperatively. At two-year follow-up,

range of motion was 0 to 114 degrees. The HSS score was 95 bilaterally. On radiologic examinations, there was no evidence of loosening of any components.

The patient was informed that data concerning the case would be submitted for publication.

Critical review of the literature

The PubMed-Medline database was searched for case reports published up to 2011. The following search string was used: (ochronosis and arthroplasty) or (ochronosis and knee arthroplasty) or (alkaptonuria and knee arthroplasty) or (ochronotic arthropathy).

Titles, abstracts and full-text publications were obtained. No language restrictions were applied. Full versions were obtained of all articles from Medline or the authors.



Figure 1. Advanced degenerative changes of the knee joints.



Figure 2. Postoperative radiographs of the knee joints: two years after operation

By this approach, we retrieved 13 articles and all were case reports of 21 knees. Cemented arthroplasty had been performed in nine, cementless in five and not reported in the remaining seven.

None of the articles defined physiotherapy in detail but all had good or excellent results (Table I).

DISCUSSION

Patients with alkaptonuria are usually asymptomatic and arthropathy appears after the fourth decade.^[3] Ochronotic arthropathy is often diagnosed intraoperatively and may not be suspected until a blackened joint is found intraoperatively.^[4,5] In the present case the diagnosis of ochronosis was not made until exploration of the joint.

Although cement is known as a risk factor for deep-vein thrombosis, according to Aslan et al.^[6] bone cement application in major joint arthroplasties such as hip or knee arthroplasties has no significant effect on the frequency of deep vein thrombosis (DVT). Konttinen et al.^[7] performed cementless type arthroplasty for the quality of bone was considered good. Additionally, Aydoğdu et al.^[8] observed that there is never any pigment associated with bone tissue itself and the bone tissue is relatively protected from the disease process and implanted cementless prosthesis, because one could expect normal bone mechanism cement-free fixation.

On the other hand, according to Fisher and Davis^[9] the bony structures may be severely affected because the case they reported had multiple fractures although they found no risk factor.

According to our observation, bony structures were not affected severely from the disease. But there is lack of experience for prosthesis type choice in ochronotic arthropathy, so we applied cemented

type prosthesis. Also our definite diagnosis was postoperative (Figure 2).

Interestingly, most of the cases in the literature had been treated by bilateral arthroplasty, and all had satisfactory results (Table I). We agree with the literature that dynamic balance parameters in the daily activities of patients with bilateral TKA were expected to be better than patients with unilateral cases.^[10] Also during the postoperative period, load asymmetry between the two extremities in patients with unilateral TKA remains the same due to advancing age. This results in the acceleration of osteoarthritis on the nonoperated side.^[11]

In ochronotic arthropathy, it is not possible to compare the results of the arthroplasty with the results of other metabolic, degenerative or inflammatory diseases.^[8] For this reason, our experience is severely limited. The solution is to perform a critical review of the literature. However, our efforts failed due to the lack of uniformity in reporting cases (Table I).

Only Aydoğdu et al.^[8] reported pre- and postoperative knee scores. So it is not clear how the other authors achieved good or excellent outcomes. In some cases, pre- and postoperative range of motion measurements were also lacking (Table I). Only Moslovac et al.^[12] gave information about type of physiotherapy. In our opinion, pre- and postoperative functional scores along with the detailed description of the physiotherapy are mandatory.

As a result we conclude that arthroplasty is a good choice for patients with ochronotic arthropathy but there is no safe information in the literature about the type of the prosthesis, so we need more experience and information. Additionally, reporting such cases in detail and uniformly may convert experience to real knowledge.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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