Irreducible cervical kyphotic deformity in a long standing case of oochronosis

A Qayum, M Vijayasaradhi, M Panigrahi

Citation

A Qayum, M Vijayasaradhi, M Panigrahi. *Irreducible cervical kyphotic deformity in a long standing case of oochronosis*. The Internet Journal of Spine Surgery. 2008 Volume 5 Number 1.

Abstract

Oochronosis is a rare clinical entity and represents pigment deposition of HGA in connectice tissue and cartilage1.The characteristic features are urine turning black on standing with blackish discoloration of sclera[figure 1], ear[figure 3], cartilage and skin. Skeletal manifestations are characterized by deposition of pigment in intervetebral disc, disc calcification [figure 2] and radiologically vacuum sign in disc space due to splitting of disc.Spinal involvement is usually in the form of spondylotic changes, but development of kyphotic deformity in normal lordotic spine is very rare. We present a case of oochronosis with cervical kyphotic deformity [figure 6] causing myelopathy.

CASE REPORT

A 49 year male patient businessman by occupation ,a known case of oochronosis diagnosed 20 years back and is under followup up of rheumatologist,presented with complaint of chronic LBA of 5 years duration,difficulty walking 4 years duration and weakness of grip of 2 years duration.He is non-diabetic non-hypertensive.He had relentless progression of symptomatology during past one year.He developed difficulty in walking during past 4 years.Patient initially felt fatigue in walking long distances,then he had difficultuy in climbing upstairs since 2 years.Presently he needs support to walk.

Patient gives h/o discoloration of urine on standing.There is h/o weakenss of upper limbs of same duration of 2 years.He is unable to write and use his right hand properly.There is no involvement of bowel and bladder.

Past history-underwent TA repair after traumatic rupture.There is h/o loose bodies in [R] knee.

Father is a known c/o Oochronosis,who died 10 years back,details not known.

ON EXAMINATION

Patient was conscious cooperative, well oriented,afebrile.pulse 80 bpm,blood pressure 130/86 mmg.no anaemia,cyanosis,jaundice,oedema ,lympadenopathy, hyprerpigmentation-sclera,ear lobules,cheeks,fingers are found[figure 1,figure 3]

Neuro exam: HF: normal, Cranial nerves Normal. Spasticity in all limbs,hyperreflexia,4/5 power in upper and 3/5 in lower limbs.sensory level C4 with graded sensory loss to all modalities of sensations.plantars bilateral extensors, gait spastic

Spine: loss of lumbar lordosis,flexion grossly restricted.SI joints normal

s/e-Chest,CVS,GI system normal

INVESTIGATIONS

Hb 12.8,PCV 31,MCV 91.8,ESR 10,ECG wnl,creatinine 1mg/dl,chest x-ray WNL,TLC 1300

DLC N82,M2,E2,blood sugar 87

CUE;PH5,specific gravity1025,sugar,albumin,ketones-nil,epith cells 3-4

Cervical x-ray:C3-7 kyphosis with kyphotic angle of 60,reduced discs paces [figure 6]

Xray L/S spine: ossified disc spaces L2-S1,with loss of lumbar lordosis with disc calcification[figure 2]

MRI co spine; cord compressionc3-7 with kypnotic deformity with obliteration of cuff spaces. No e/o myelomalacia

Figure 1

Figure 1

Figure 2

Figure 2

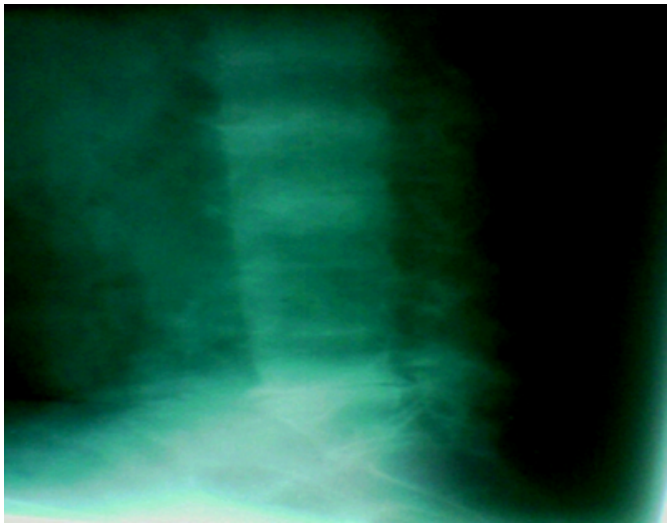


Figure 3

Figure 3

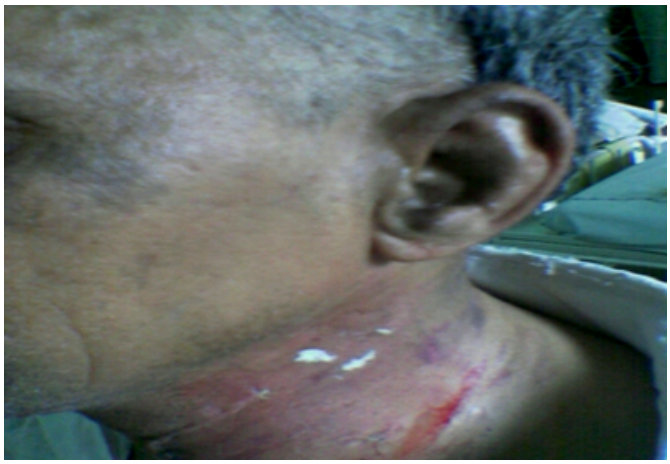


Figure 4

Figure 4

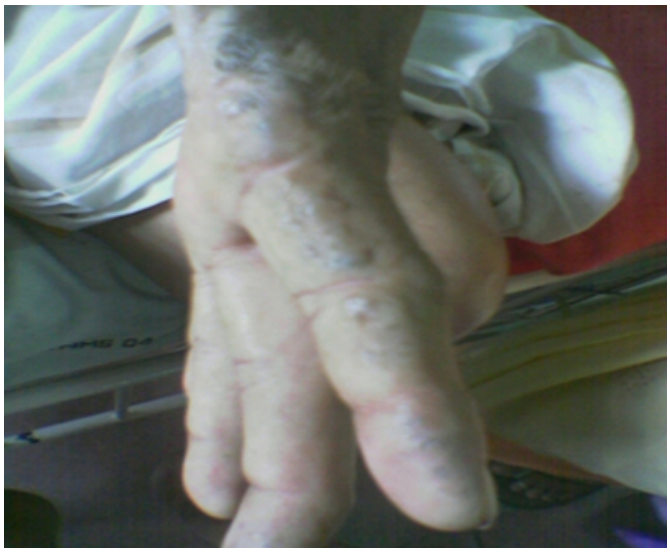


Figure 5

Figure 5

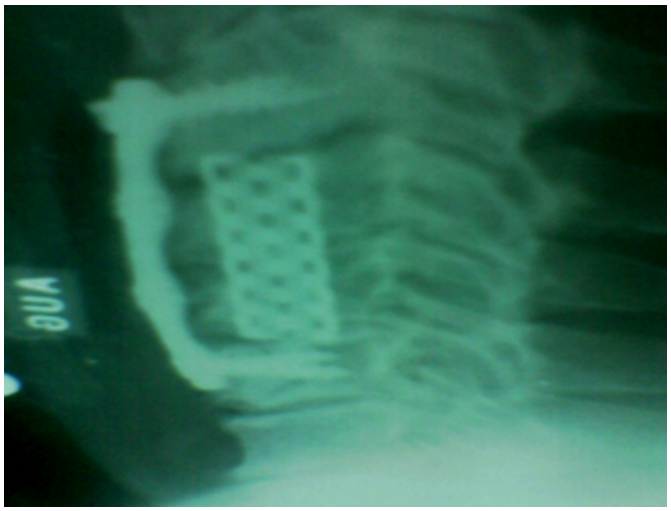
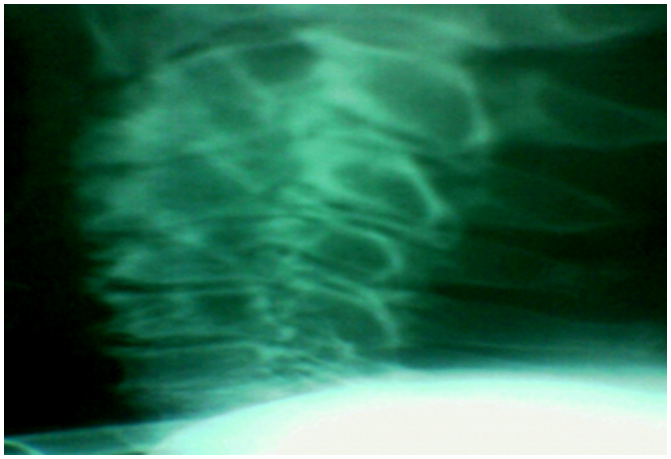


Figure 6

Figure 6



TREATMENT

ACDF was done.

TECHNIQUE

Routine anterior cervical approach under GA with bronchoscopic intubation. C4 and C5 corpectomy with spacer and plate fixation[figure 5] was performed with adequate decompression using high speed drill.Intra operative maneuvers to reduces deformity were not successful due to very stiff spine.

Preoperatively prevertebral fascia,disc spaces and disc material were black. Procedure was uneventful and patient was discharged on 5th POD.postop x-ray shows spacer in place and no reduction of deformity [Figure 5]patient improved in spasticity and power in lower limbs improved by grade 1.patient is being follwed by physiotherapist.

DISCUSSION

Alkaptonuria is a rare disease with incidence of 1;1 milllion [3].The disease was first described by Scribonius in 1854 in a child whose urine was black. Thereafter Boedeker and Wirchow reported a few cases in 1859 and 1866,respectively.Albrecht and Zdareck called attention to the association between alkaptonuria and oochronosis in 1902.In this disease,the first symptom related to joint involvement are generally back stiffness and low back pain. The molecule occurring with oxidation and polymerization of HGA binds to collagen irreversibily[3].Accumulation of this molecule in the cartilage of joints and IVD causes degradation of cartilage.

In alkaptonuric spondylosis,degenerative changes may be seen along the whole of he spine,however most prominent involvement is in lumbar region.Initially there is loss of lumbar lordosis,thoracic kyphosis becomes prominent.So,radiologically it mimicks ankylosing spondylitis.There are no syndesmosis,annular ossification or bamboo sign in alkaptonuria. Progressive degeneration and calcification of NP causes characteristic radiographic finding described by Pomeranz te al as universal calcification of Intervertebral disc 5.Calcification appeared a elliptical opaque wafers .Vacuum phenomenon has often been reported.Involvement of cervical spine is late[6]

Kyphotic deformity of cervical spine is a very rare entity and pathogenesis is unclear.once develops,it progresses,hence need for screening of patients .once deformity occurs,correction is very difficult due to extensive calcification and stiff spine.Hence aim of surgery is to reverse neurodeficits and prevent further deformity.

CORRESPONDENCE TO

Dr Abdul Qayum
M.B,B.S,M.S[ortho][Kashmir university]
F.S.S[spine fellowship][NIMS-Hyderabad]
Bangri Palace,Dr Guru’s lane
Barzulla Baghat,Srinagar Kashmir
Jammu & Kashmir[india]
- 190005
Ph: 9797809141

References

1. Bayindir,Petek,yilmaz Ovali,Gulgun,Pabuscu,Yulkel,TemizCuneyet,Durouz,Tuncay:radiological features of lumbar spine in oochronosis in late stages:clinical rheumatology,vol 25,number 4,july 2006,pp 588-590
2.

Balaban,Birol;taskaynatan,mehmet,yasar,evren,tan,kenan,kalyon,tunc:ochronotic spondyloarthropathy:spinal involvement resembling AS:clinical Rheumatology,volume 25,No 4,july 2006,pp 598-601
3. Millea TP,segal LS,Liss RG,stauffer ES,spine fractures in oochronosis:Clin Orthop Relat Res.1992 Aug9281-11
4. Field Jr,Hgely GB,De saussure RL.Oochronosis with lumbar Disc;Case report.J Neurosurg 1963;20:348-51
5. Pomeranz MM,Friedman LJ,Tunick IS.Roentgen findings in alcaptonuric oochronosis.Radology 1941;37:295-302
6. Sharma RR ,Ravi,Bhama BA,et al.cervicomedullary compression secondary to ochronotic arthritis affecting craniovertebral joints syndesmo-dental ochronotic arthritis. J Postgrad Med 1988;34:253-7

Author Information

Abdul Qayum, M.S.
Department of neurosurgery, Nizam's institute of medical sciences

Mudumba Vijayasaradhi, MCh
Department of neurosurgery, Nizam's institute of medical sciences

Manas Kumar Panigrahi, MCh
Department of neurosurgery, Nizam's institute of medical sciences