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GRAND ROUNDS

Management of a pseudarthrosis with sagittal malalignment in a patient with ochronotic spondyloarthropathy

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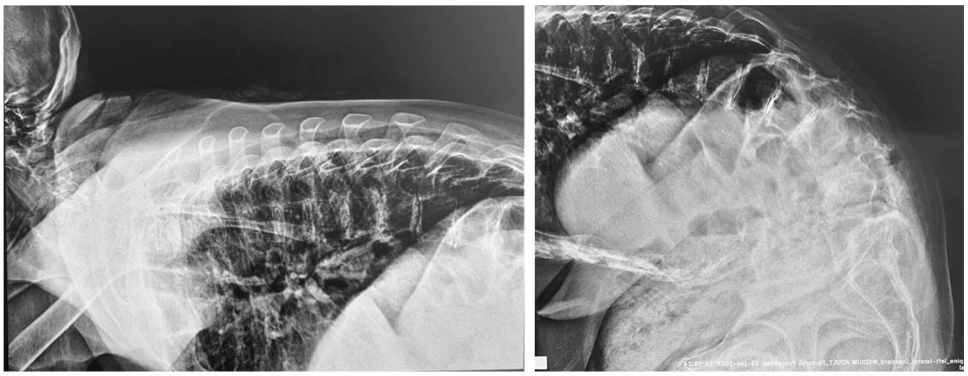
Abstract

Purpose Ochronotic spondyloarthropathy is an uncommon disease, and its association to sagittal malalignment in the context of a pseudarthrosis has never been described.
Methods We present the case of a 56-year-old female, who underwent previously L4L5 laminectomy for central canal stenosis and started later on to complain of progressively severe low back pain with a significant forward imbalance while walking. X-rays showed non-compensated sagittal malalignment due to thoracolumbar kyphosis, CT scan revealed multilevel central intradiscal calcifications with important vacuum disc at L4L5, and MRI showed T1 and T2 hypointensity signal at the same level with bone marrow oedema. Alkaptonuric ochronosis was suspected and confirmed by the presence of homogentisic acid in the urine, and the diagnosis of L4L5 pseudarthrosis with associated severe sagittal malalignment in the context of ochronotic spondyloarthropathy was established.
Results The patient underwent surgery with a posterior-only approach with a long-segment pedicle screw construct from T10 to the pelvis with a 360° fusion with a cage at L4L5. Samples taken from the disc and ligaments confirmed the diagnosis of ochronotic spondyloarthropathy macroscopically and microscopically. She could walk on day 2 with a satisfactory clinical and radiological result at 2 years.
Conclusion This is the first case in the literature to describe a post-laminectomy pseudarthrosis leading to a significant sagittal malalignment in a patient with ochronotic spondyloarthropathy. Management of such a case is challenging as the spine is partially ankylosed; therefore, a long construct is advisable to avoid ankylosing disorders related complications.

Keywords Alkaptonuria · Ochronotic spondyloarthropathy · Pseudarthrosis · Sagittal alignment · Kyphosis

Extended author information available on the last page of the article



(a) (b)

Fig. 1 Standing full spine X-ray lateral view divided into two images as one image could not be obtained because of the severe sagittal malalignment (**a**, **b**)

Case presentation

A 56-year-old female has been complaining of progressive forward imbalance during the past 4 years.

Her past history revealed generalized joint pain since early adulthood with chronic low back pain and progressive restriction of the movements of the thoracic and lumbar spine in the last 10 years. She underwent 6 years before, in another city, L4L5 laminectomy with partial facetectomies on both sides for neurogenic claudication due to central canal stenosis and started 2 years later to complain of progressively disabling low back pain with a significant forward imbalance while walking, requiring anterior support (cane). Physical examination revealed a patient spontaneously in a forward bending position, with impossibility to stand up straight, painful in supine position with impossibility to lie completely flat, but normal neurological findings.

On the X-rays, the L4L5 intervertebral disc space was widely opened, with major vacuum phenomena and sclerotic margins. CT scan confirmed the radiological aspect with multiple levels disc calcification. Sagittal MRI showed hypointensity of the L4L5 anterior defect on the T1 and T2 weighted sequences with bone marrow oedema.

After discussion with the rheumatologist, alkaptonuric ochronosis was suspected and confirmed by the presence of homogentisic acid in the urine.

The diagnosis of pseudarthrosis of the L4L5 level with associated severe sagittal malalignment in the context of ochronotic spondyloarthropathy was then established, and given the fact that the patient already exhausted during the previous months, all non-operative therapies including medications, physical therapy and injections, and surgical intervention was proposed as a single-stage posterior correction with 360° fusion at the pseudarthrosis level and a long construct from T10 to the pelvis.

Diagnostic imaging section

Due to the severe forward imbalance, proper standing full spine X-rays could not be obtained and the lateral view was divided into two images, revealing a significant sagittal malalignment with very positive sagittal vertical axis and thoracolumbar kyphosis (Fig. 1a, b). Supine lateral and anteroposterior thoracolumbar X-rays showed partial reduction of the kyphosis with reduced intervertebral disc spaces, with calcification within the discs (Fig. 2a), and a vacuum phenomenon at L4L5 (Fig. 2b). Computed tomography scan confirmed the intervertebral discs calcification with important vacuum disc at L4L5 with sclerotic endplates, previous laminectomy at L4L5 and pseudoblocks at L3L4 and L5S1 (Fig. 3a–c). Magnetic resonance imaging of the thoracolumbar spine showed loss of intervertebral disc height with multiple level central intradiscal low signal on T1 and T2 that is consistent with disc calcification, also the L4L5 anterior defect showed a hypointensity signal on the T1 and T2 weighted sequences with hyperintensity of the adjacent vertebral bodies on FAT SAT attesting bone marrow oedema (Fig. 4a–c).

Historical review, epidemiology, diagnosis, pathology and differential diagnosis

Alkaptonuria is a rare autosomal recessive disorder with an incidence of 1 in 250,000 to 1 in 1,000,000 live births [1]. It is caused by an enzyme deficiency leading to abnormally high levels of homogentisic acid in blood and urine, which is an intermediate product in the metabolism of tyrosine. Excess amount of homogentisic acid gets oxidized and is deposited in the articular cartilage as ochre-coloured pigment granules, and as it accumulates, the cartilage loses elasticity and becomes brittle, leading to calcification and eventual break down [2]. Pathophysiology describes four progressive stages: inflammatory (stage 1), early disc

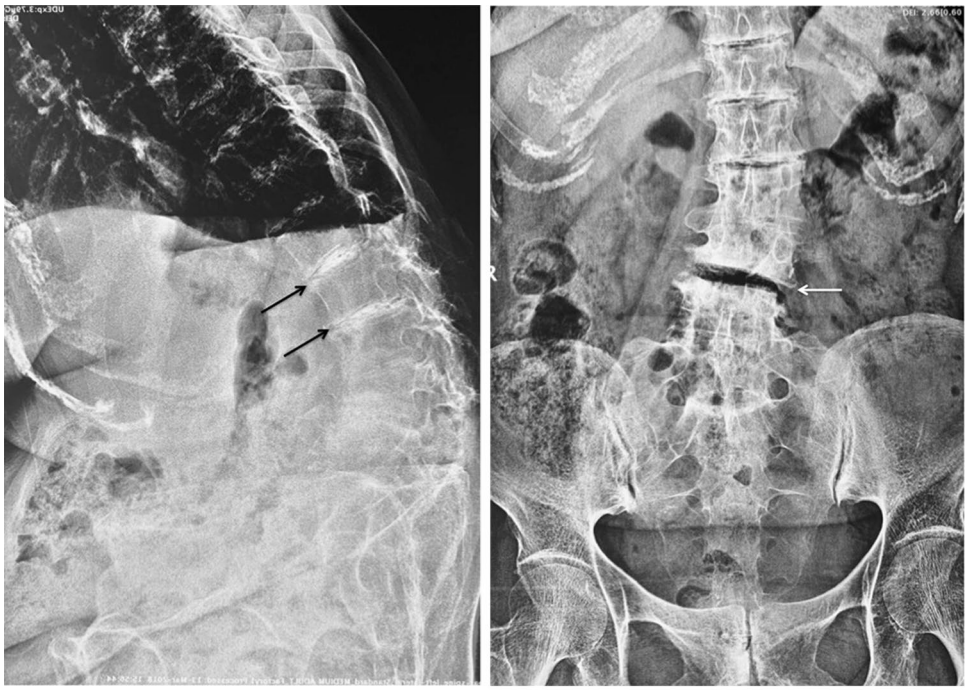


Fig. 2 Supine lateral and anteroposterior thoracolumbar X-rays showing partial reduction of the kyphosis with calcification within the discs (**a**, black arrows), and a vacuum phenomenon at L4L5 (**b**, white arrow)

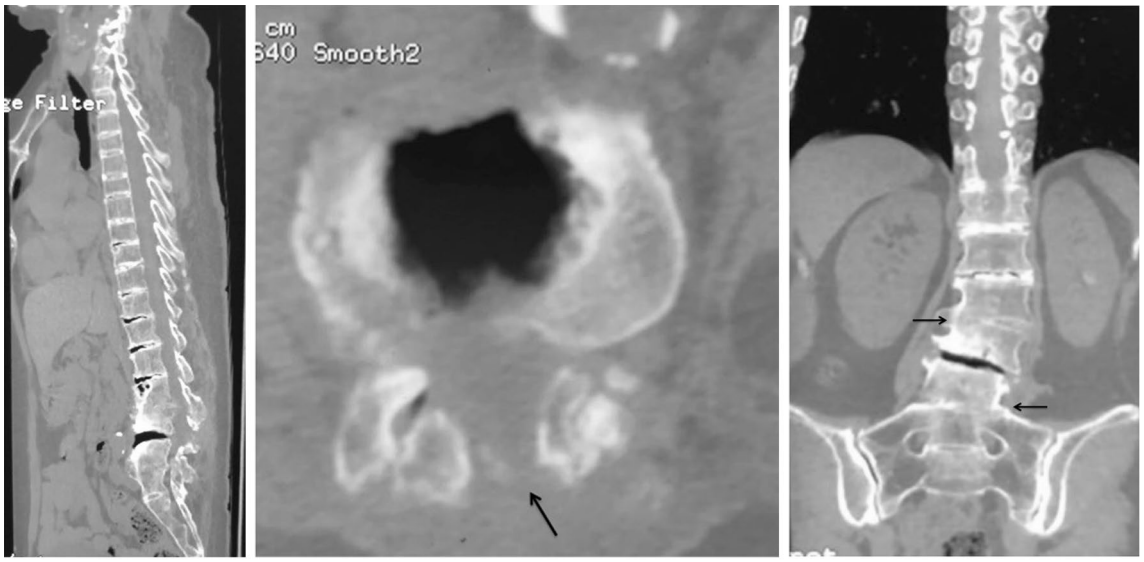


Fig. 3 CT scan confirming the multiple thoracolumbar intervertebral discs calcification with vacuum phenomena mainly at L4L5 with sclerotic endplates (**a**), previous laminectomy at L4L5 (**b**, black arrow) and pseudoblocks at L3L4 and L5S1 (**c**, black arrows)

calcification (stage 2), fibrous ankylosis (stage 3) and bony ankylosis (stage 4) which maybe confused with ankylosing spondylitis [3, 4]. Because of its rarity, the literature is scarce about ochronotic spondyloarthropathy and it has been described only as case reports [5–8] with no big series. Characteristic features of ochronosis include blue dark discolouration of connective tissues including sclera, cornea, heart valve, articular cartilage, tendons and ligaments [9]. Thus, clinical presentation may reveal grayish pigmentation of the sclera or the ear cartilage, with spinal movements restriction, and urine turns dark brown on exposure to air, but it is usually asymptomatic until the third or fourth decade. X-ray and CT scan usually show intervertebral discs calcifications with severe degenerative changes, that may

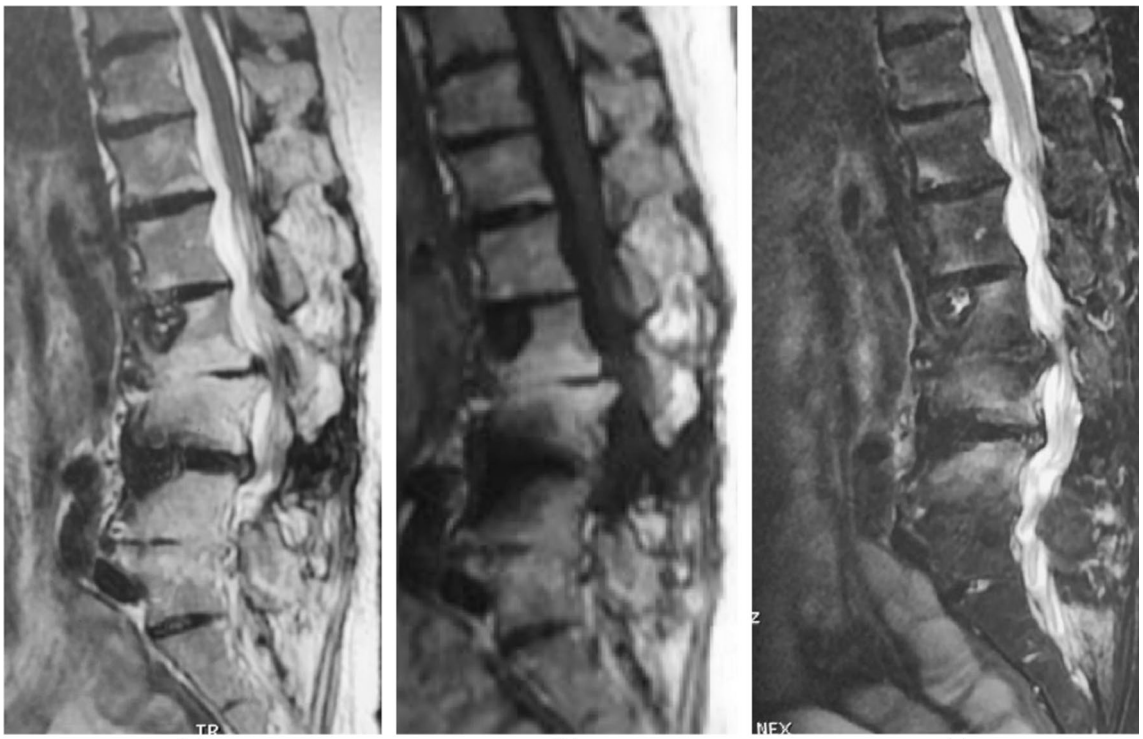


Fig. 4 MRI showing multiple level central intradiscal low signal on T1 and T2 weighted sequences consistent with disc calcification, and the L4L5 anterior defect in hypointensity signal on T1 and T2 (**a**, **b**) with bone marrow oedema of the adjacent bodies on FAT SAT (**c**)

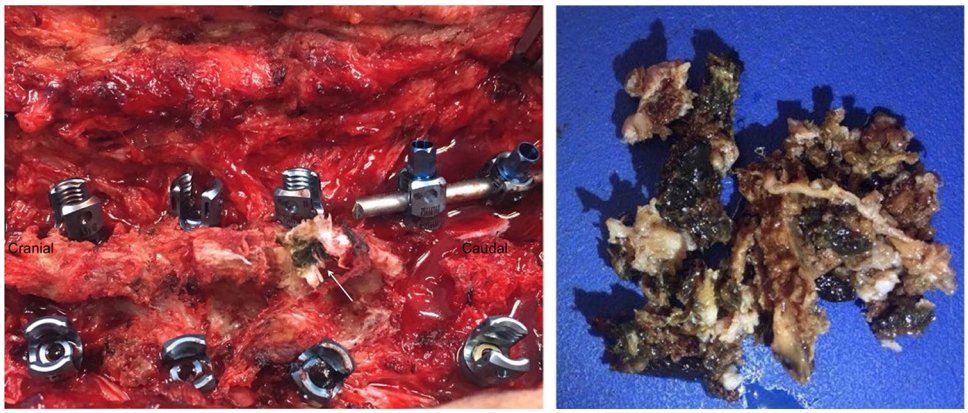
lead to a disc protrusion [10, 11], thoracic myelopathy [7], rarely pseudarthrosis in the lumbar or thoracic spine may occur [12]. Magnetic resonance imaging reveals low disc height with low signal within the disc on the T1 and T2 weighted sequences, and in case of pseudarthrosis, the affected disc is demonstrated as an irregular lesion with either hypointensity T1 and T2 or hypointensity T1 with hyperintensity T2 [12]. Diagnosis can be confirmed by the presence of homogentisic acid in the urine. In the case of our patient, initial laminectomy with partial facetectomies from the previous surgery created a hypermobile L4L5 level between two blocks above and below and was the key factor in the occurrence of pseudarthrosis that progressively lead to a significant sagittal malalignment.

Rationale for treatment

Currently, there is no specific and effective treatment available for ochronosis, therefore, and as a global management, symptomatic and supportive therapies are necessary to prevent further disability.

Pseudarthrosis of the lumbar spine with sagittal malalignment in ochronotic spondyloarthropathy is rare, and no optimal management options have yet been established. Exhaustion of non-operative therapies such as medications, physical therapy, injections and immobilization in a brace [12] should be done before proceeding to any type of surgical intervention. Surgical management of such lesion can be done either by an anterior approach or a posterior approach, or both. The combined approach consisting of an anterior debridement of all necrotic tissues from the gap with placement of a cage or a shaped autograft, followed by a posterior instrumentation is a well-described surgical option in the treatment of pseudarthrosis in all ankylosing conditions [13, 14]. The anterior-only approach has the disadvantage of not fixing the vertebra above and below the pseudarthrosis posteriorly; therefore, non-union was reported in the literature [15]. The posterior-only approach has gain in popularity during the past decade as it enables achieving a 360° fusion from the back, in association with a posterior instrumentation with good correction of any eventual associated sagittal malalignment; it avoids an additional anterior approach, thus decreasing operative time, blood loss and hospital stay [12]. Long-segment instrumentation has been advocated in the case of pseudarthrosis or fracture in ankylosing spinal disorders [12, 16] as it guarantees the alignment and stability on the long term.

In the case of our patient, given the sagittal deformity with L4L5 instability, multiple discs disease with vacuum



(a) (b)

Fig. 5 Peroperatively, the interspinous ligament revealed a black pigmentation (**a**, white arrow) and the removed disc material was found to be pigmented in black and brown (**b**)

phenomenon up to the thoracolumbar area and several ankylosed levels, a long-segment pedicle screw construct from T10 to the pelvis with insertion of a high cage in L4L5 by the mean of a transforaminal approach was done.

Operative procedure

Under general anesthesia, with a patient in a prone position, on four cushions, spinal cord monitoring was used in the form of transcranial motor evoked potentials, somatosensory evoked potentials and free running electromyography (EMG) of the lower extremities and evoked EMGs with pedicle screw stimulation. A posterior cutaneous midline incision was made. The spine was exposed subperiosteally from T10 to S2, in a fashion similar to other posterior instrumented surgeries, going laterally to the transverse processes and sacral ala, with care taken to preserve the supraspinous and interspinous ligaments proximally between T9 and T10. Inferior facetectomies were performed to provide maximum flexibility, the spinous processes were also resected, and the bone recovered was prepared for use as a graft at the end of the procedure. Free-hand technique was used to place the pedicle screws at all levels, from T11 to the sacrum with insertion of S2 alar iliac screws under image guidance. Proximally, hooks were inserted at the level of T10 to enable a smoother transition with the junctional area cranially. The interspinous ligament that was removed revealed a black pigmentation (Fig. 5a), which was macroscopically in favour of ochronosis.

Ponte osteotomies were then performed at the relatively flexible levels L1L2, L2L3 and L4L5, and at the latter level, both foramens were widely opened, laterally to the fibrosis from the previous surgery, and this enabled complete removal of the disc on both sides which showed brown-dark colouration (Fig. 5b), curettage of the sclerotic endplates and insertion of a high cage (14 mm) from the left side, with complementary autologous bone graft inserted from the right side.

Two 5.5 mm cobalt chrome rods were secured distally and gradually cantilevered to the proximal screws for progressive reduction of the kyphosis, additional satellite rods were added to strengthen the construct around L4L5, and this was completed by compression manoeuvres at the Ponte osteotomy levels to increase the correction.

Posterior elements were decorticated using a high-speed burr, and bone graft (autologous and allograft) was placed to cover the maximum surface. Disc samples were sent to the laboratory for histopathological examination.

Clinical outcome

The patient started to walk on day 2 with the assistance of a walking frame and was discharged on day 6. She showed a significant clinical improvement of her sagittal malalignment. Pathological specimen showed pigmented disc tissue embedded in granulation tissue, thus confirming ochronosis, and postoperative thoracolumbar X-ray confirmed the good position of the cage in L4L5 with correction of the thoracolumbar kyphosis (Fig. 6). She showed a satisfactory clinical and radiological result at 2 years (Figs. 7a, b, 8).

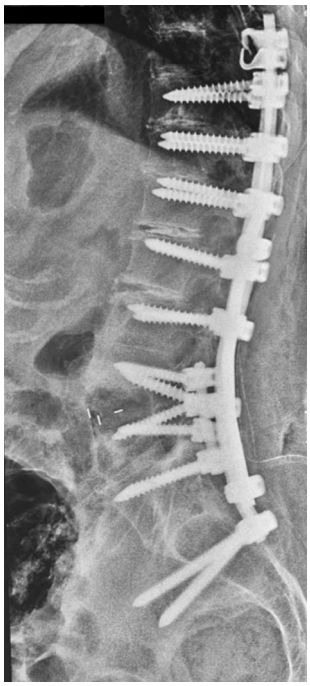


Fig. 6 Immediate postoperative thoracolumbar X-ray confirming the good position of the cage in L4L5 with correction of the thoracolumbar kyphosis

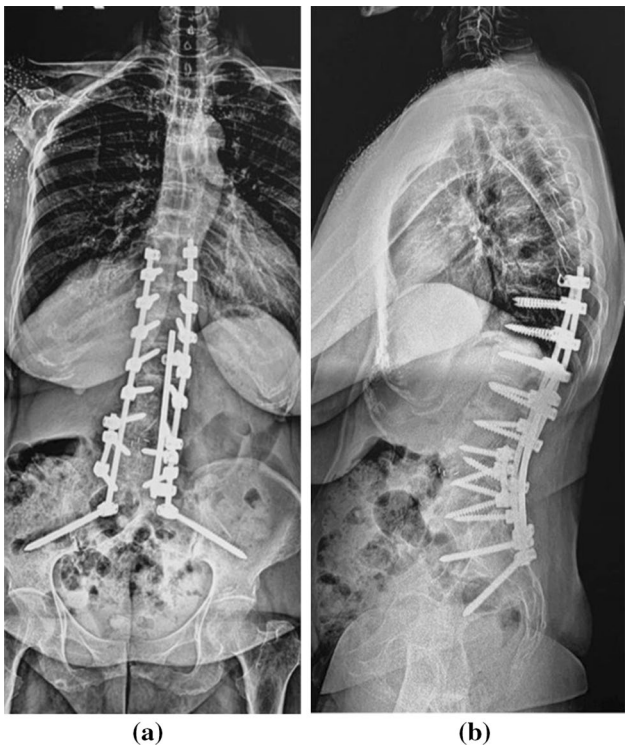


Fig. 7 Full spine X-rays 2 years after the surgery showing a satisfying alignment with a stable construct

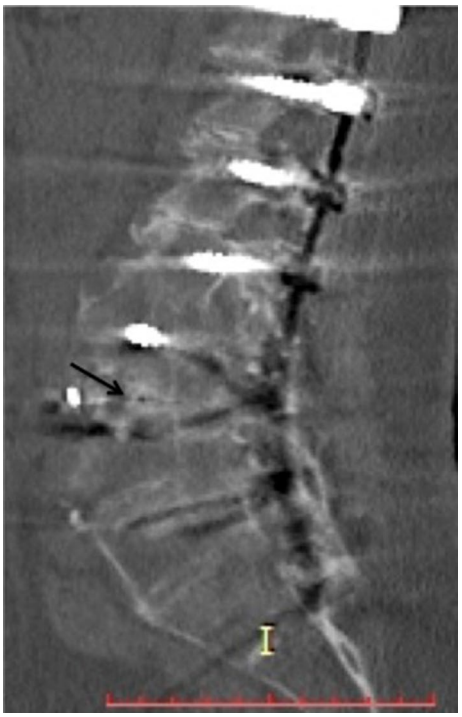


Fig. 8 CT scan at 2 years confirming the fusion at L4L5 (black arrow)

Compliance with ethical standards

Conflict of interest There are no conflicts of interest for this case report.

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