CASE REPORTS AND TECHNICAL NOTE

OCHRONOSIS ASSOCIATED WITH DEGENERATION OF AN INTERVERTEBRAL DISC

ROBERT G. FISHER, M.D., AND JOHN WILLIAMS, M.D.

_Hitchcock Clinic, Dartmouth Medical School, and Hitchcock Hospital, Hanover, New Hampshire_

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CASE REPORT

J. G., a 32-year-old white male, was admitted to the Hitchcock Hospital on Oct. 14, 1954 with the diagnosis of a protruded intervertebral lumbar disc.

Six weeks prior to admission, the patient lifted a loaded milk can on to the rear of a pickup truck and had immediate stabbing pain in the back. He had been in excellent health previously. He went to a chiropractor who relieved the pain in his back but caused severe pain in the right leg. There were no pains in the left leg and no disturbances of bladder. The pain in the right leg was made worse by coughing and sneezing. There were paresthesias in the right foot and toes.

A system review was negative save that he had been studied 8 years previously for aches of bones and joints and was thought to have had a “touch of polio.” He was married and the father of three children. He had been in the U. S. Army for 3 years and had been in the Pacific Theater in World War II. He used no drugs. One older brother had had backaches for a long period of time but these had been attributed to “arthritis.”

Examination. General physical findings were normal. His posture was normal but flexion of the spine in the lumbar region was limited to 10°. Walking on heels and toes was normal. There was severe spasm of the lumbar muscles. The right ankle jerk was absent. The other deep reflexes were normal. Motor and sensory findings were normal bilaterally. Babinski responses were plantar bilaterally. Straight leg raising on the right side was limited to 30° and there was a positive Lasègue sign on this side. The findings in the left leg were normal.

Red and white blood cell counts and differential white cell count were normal, and the Mazzini reaction (syphilis) was negative. The initial urinalysis showed a slightly positive reaction for sugar but no acetone or diacetic acid; two later urinalyses gave normal findings. No abnormality of color was noted. Roentgenograms of the chest and spine were negative except for narrowing of the lumbosacral space. A myelogram done elsewhere disclosed a defect at the lumbosacral space on the right side.

Operation. On Oct. 15, 1954 the patient underwent a laminotomy at the right lumbosacral space under spinal anesthesia. The laminae and the yellow ligament were normal. The posterior longitudinal ligament was markedly elevated and pressing on the 1st sacral root; the ligament was white in appearance. An incision into the ligament disclosed a black-appearing disc which was degenerated and soft (Fig. 1). This was easily removed in large pieces. A partial laminectomy at the 1st sacral segment was done and loose black cartilaginous material was found under the root, lying free in the canal. The root was well decompressed and closure was accomplished uneventfully.

Postoperative Course. There was transient inability to void. An indwelling catheter was utilized for 3 days. The urine was noted to be black upon standing, and reduced Benedict’s solution. Ochronosis was established as the diagnosis. Closer examination of the patient failed to show any signs of the disease except for minor discoloration of the ears. Extensive roentgenographic investigation of the lumbosacral spine, hips, shoulders, hands and knees disclosed no abnormality.
He was free from pain after the 2nd postoperative day. He was placed on 100 mg. of ascorbic acid and citrus flavinoid compound three times daily although we did not anticipate any change in the color of the urine. He was given a course of strengthening exercises and was discharged on the 12th postoperative day. He remained symptom-free although he has recently passed a kidney stone and this problem is under investigation.

His older brother was investigated and found to have alcaptonuria and roentgenographic changes typical of ochronosis of the spine. The cartilages of the ears, nose and sternum were very black and his perspiration was dark-colored and stained his clothes. He was bothered by pain in the lumbodorsal region of the spine. The disease was not found in other siblings or children.

**DISCUSSION**

Ochronosis is a term used by Virchow in 1866 to designate a condition in which the cartilages of the body appear black rather than white. The microscopic appearance of the granules in the cartilages was ochre rather than black. No mention was made in the article about the urine appearing black on exposure to the air; but Boedeker in 1858 first recognized a substance in urine which could combine with oxygen in an alkaline medium to form a substance which he termed “Alcapton.” The term alcaptonuria was derived from this. Wolkow and Baumann in 1891 first recognized this compound as homogentisic acid. This compound appears in the urine when either of the two essential amino acids, phenylalanine and tyrosine, is incompletely metabolized. Homogentisic acid also appears in the urine of those having phenylpyruvic oligophrenia, tyrosinosis, or ascorbic acid deficiency.

Homogentisic acid does not discolor acid urine but causes the urine to appear black or brown if alkaline. It reduces Benedict’s solution, does not ferment yeast, and does not rotate polarized light. This enables one to differentiate it from glucose and thus avoid confusion with diabetes mellitus. The occurrence of this compound is attributed to failure of the enzyme system to convert homogentisic acid into fumarylacetooacetic acid and eventually to fumaric acid and acetooacetic acid.

Virchow found the ochre pigment deposited in the eyes, skin, and hands, as well as the cartilages of the feet, ribs and large joints. It has been identified in many