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Case Report

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Paget's disease of bone presented as normal pressure hydrocephalus: A case report and review of literature

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Keywords
Dementia, Paget's Disease, Normal Pressure Hydrocephalus

Abstract

Background: Paget's disease is a focal bone disorder manifested as bone overgrowth and disrupted bone integrity as a result of accelerated bone remodelling rate. Rarely, Paget's disease of the base of the skull results in hydrocephalic dementia, and the triad of normal pressure hydrocephalus syndrome is a much more scarce entity.

Case Report: Herein, we report an elderly woman who presented in Imam Khomeini Hospital, Tehran, Iran, with normal pressure hydrocephalus syndrome due to Paget's bone disease. Furthermore, we have reviewed relevant previous studies

Conclusion: Paget's disease can be presented as normal pressure hydrocephalus syndrome.

Introduction

Paget's disease of bone is considered as a focal disorder of bone metabolism, manifested as bone overgrowth and disrupted bone integrity as a result of accelerated bone remodeling rate.[1] This disorder is mainly asymptomatic and the overall age- and sex-standardized prevalence is estimated around 0.3%.[2] To date, the etiology of Paget's disease remains unknown. Genetic factors and viral infections are postulated as possible etiologies.[3] The major manifestations of Paget's disease of bone include pain, pathologic fractures, and skeletal deformities, especially at long bones, skull, and clavicles.[4] Furthermore, nerve entrapment by bone overgrowths results in diverse neurologic manifestations.[4] The disease is suspected when an isolated elevated serum alkaline phosphatase with normal calcium and phosphorus levels is observed. Bone scan is considered as the most sensitive diagnostic test in identifying pagetic bone lesions.[4]

Normal pressure hydrocephalus refers to enlarged ventricles with normal opening pressure on lumbar puncture. It is manifested with dementia, gait disturbance, and urinary incontinence. The manifestations are reversible using ventriculoperitoneal shunt.[5]

In the current literature, hydrocephalus remains as a rare complication among patients with Paget's disease. Hence, the presenting manifestation as normal pressure hydrocephalus is a much more scarce entity. There is a scarcity of normal pressure hydrocephalus as presenting manifestation of Paget's disease. Therefore, we report a 66-year-old female presented to Imam Khomeini Hospital, Tehran, Iran, with the chief complaint of urinary incontinence, gait abnormalities, and memory deficits. The brain CT scan revealed hydrocephalus in all supratentorial ventricles.

Case Report

A 66-year-old woman was admitted to Imam Khomeini Hospital with the chief complaint of gait difficulty since

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6 years ago. She described the gait difficulty as a falling tendency while walking. She also complained of urinary incontinence. Furthermore, she expressed annoying memory problems. She had diabetes type II, hypertension, dyslipidemia, and hypothyroidism in past medical history. Moreover, she had ischemic heart disease and a previous angiography had revealed stenosis, which was treated by percutaneous coronary intervention. The drug history comprised the following medications: amlodipine, losartan, atenolol, nitrocontin, glibenclamide, atorvastatin, metformin, and levothyroxine. Her pulse rate was 90 per minute with blood pressure of 140/90. Her recent memory was disrupted. A slight facial paresis was detectable as asymmetric nasolabial fissure pattern. She also utilized hearing aids due to hearing problems. Assessing the cerebellar function, the finger to nose and heel to shin tests were normal, while the tandem gait was disrupted. Her gait was slow but normal for her age.

Cell blood count showed lymphocytosis of 42.1% with the white blood cell (WBC) count of 5500/µl. The brain CT scan revealed thickened calvaria (Figure 1). Furthermore, hydrocephaly was detectable in all supratentorial ventricles, but the forth ventricle remained intact (Figure 2),. Besides, emphysema was observable in all subcutaneous areas around the skull and outside calvaria (Figure 1). MRI showed periventricular leukoencephalopathy and increased marrow signal (Figure 2). In order to assess the thickened calvaria, whole body scan was performed and the serum levels of calcium, phosphorous, and alkaline phosphatase were assessed.

In order to investigate the problem, cerebral spinal fluid (CSF) sample was collected after lumbar puncture. After the lumbar puncture the patient's problems including recent memory deficits diminished for one day. Venereal disease research laboratory test (VDRL) and Wright test of CSF were negative. Other indicators of the CSF sample were normal. To investigate multiple myeloma, serum and urine proteins electrophoresis failed to show any specific abnormality. The serum B12 level was normal. Serum calcium and phosphorous levels were within normal limits. Serum alkaline phosphatase was 4723IU/L. The symptoms relieved dramatically after lumbar puncture.

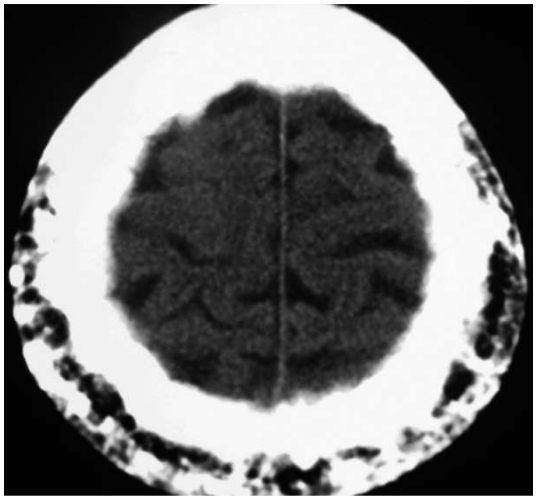

Figure1. Thick skull in brain CT scan

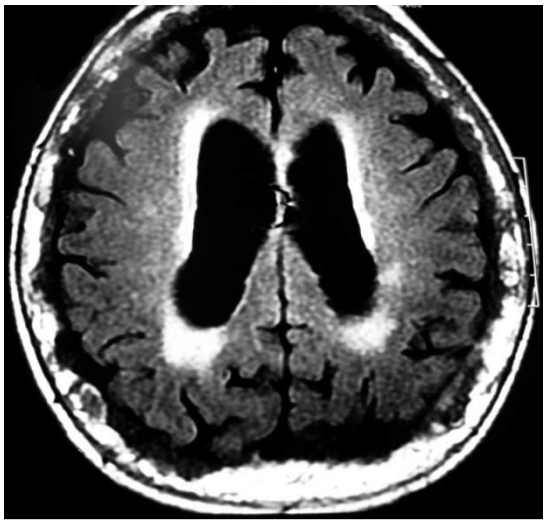

Figure 2. Hydrocephalus with interstitial edema and increased bone marrow signal in T2 weighted MRI

Discussion

Paget's disease of bone is considered as a disorder in bone metabolism which results in diverse neurologic symptoms. Pain is considered as one of the major symptoms which may arise from pagetic lesions, or may be the result of possible complications in adjacent tissues, such as arthritis or nerve impingemen.[4] our patient did not mention any pain. Other possible clinical manifestations included skeletal deformities and pathologic fractures.[4] Diverse neurologic complications were considered as another major clinical presentation.[4] These symptoms are mostly caused by nerve entrapment as a result of enlarged bone, or as an outcome of diminished blood supply. Our patient suffered from hearing loss which is recognized as a common presentation in about 37% of patients and is mainly caused by the compression of the 8th nerve. Moreover, the patient presented with facial palsy as a result of the compression of the 7th nerve.

The most prominent clinical feature in our patient was hydrocephalus. Hydrocephalus stems from the involvement of the skull base and the resultant blockage of the sylvius aqueduct. The major initial presentation in our patient, at the time diagnosis, was dementia. After lumbar puncture, the recent memory deficit improved dramatically. Hence, the patient became a candidate for shunt administration. Other

accompanied manifestations included gait abnormality and urinary incontinence. The manifestations were compatible with normal pressure hydrocephalus.

Previously, there have been few reports of Paget's disease and associated normal pressure hydrocephalus. Gottschalk was the first to report the mentioned manifestations in 1973.[6] Hens and van den Bergh reported a patient with the triad of dementia, urinary incontinence, and gait abnormality accompanied with obstructive hydrocephalu.[7] Goldhammer et al. reported an elderly woman with ataxia and dementia, related to hydrocephalus, resulting from Paget's disease of the skull.[8] Lobato et al. described a syndrome resembling normal pressure hydrocephalus due to Paget's bone disease.[9] Aguiar et al. reported an elderly woman with normal pressure hydrocephaly due to Paget's disease in 1994.[10] In 2000 Chan et al. reported an elderly woman with the triad of normal pressure hydrocephalus presented 3 years after the diagnosis of the Paget's disease mainly affecting the skull.[11] Roohi et al. reported a patient of normal pressure hydrocephalus, resulting from Paget's disease of the skull base in 2005. The reported patient was symptom free during the 6 years after a ventricular shunt was inserted.[12]

In conclusion, Paget's disease and NPH could accompany each other, and hydrocephalus symptoms may unmask underlying Paget's disease.

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