Paget's disease presenting with multiple cranial neuropathies

Srinath Vasagiri[1], Anuja Patil[2], Praveen Kulkarni[3]

From [1]Resident, [2]Consultant, Department of Neurology, [3]Consultant, Department of Medicine, Krishna Institute of Medical Sciences, Hyderabad, Telangana, India

A 67-year-old gentleman presented with complaints of new-onset right eye ptosis, dysarthria with nasal intonation of voice, and progressive dysphagia over 10 days. About 2 weeks before the onset of these symptoms, he had suffered accidental trauma to his left leg, followed by local tenderness and swelling. The patient was diabetic for the past 4 years and is compliant with treatment. On enquiring, he revealed a diminution of hearing on the right side over the past 2 months. On examination, he was afebrile, with a blood pressure of 130/80 mmHg, pulse rate of 84 bpm, and saturation of 99% at room air. Neurological examination revealed right eye partial ptosis with preserved other extraocular movements, right facial lower motor paresis, bilateral sensorineural hearing impairment, and asymmetric soft palate movements with the right glossopharyngeal nerve impairment. Cognition, sensory examination, power testing for the axial and appendicular muscles, and deep tendon reflexes were normal. The left foot showed signs of local induration and tenderness suggestive of cellulitis. In view of unilateral predominant cranial neuropathies, differential etiologies involving the extra-axial path of the contiguous nerves, including pachymeningitis and skull base osteomyelitis apart from neuromuscular junction abnormalities, were considered.

Nerve conduction studies revealed normal study including negative repetitive nerve stimulation. However, serum alkaline phosphatase was elevated to 1387 units/L (N: 56–119 units/L). The rest of the biochemical examination, hormonal profile, and tumor markers including prostatic specific antigen levels were normal. Computed tomography brain showed highly enlarged diploic spaces with intermixed sclerotic and lytic tissue areas (Fig. 1c). Magnetic resonance imaging brain revealed diffuse right calvarial expansion, trabecular coarsening with heterogenous T1 and T2 signal, and patchy enhancement with contrast (Fig. 1a and b). This is characteristic of the mixed phase between the osteolytic and osteosclerotic phases of evolution in Paget's disease [1]. Three phase bone scan showed expansile lesions with increased perfusion and osteoblastic activity right calvarium, scapula, sacrum, left hemipelvis, and bilateral proximal femori (Fig. 2). D12 vertebra showed a typical "mouse face" appearance [2] with increased uptake in the body, posterior elements, and spinous process suggestive of Paget's disease. No tracer uptake was seen in the left foot with cellulitis changes. He was treated with amoxicillin and local wound care for leg cellulitis. He underwent debridement for the left leg cellulitis after 2 weeks of antibiotics course. He was treated with risedronate 35 mg weekly

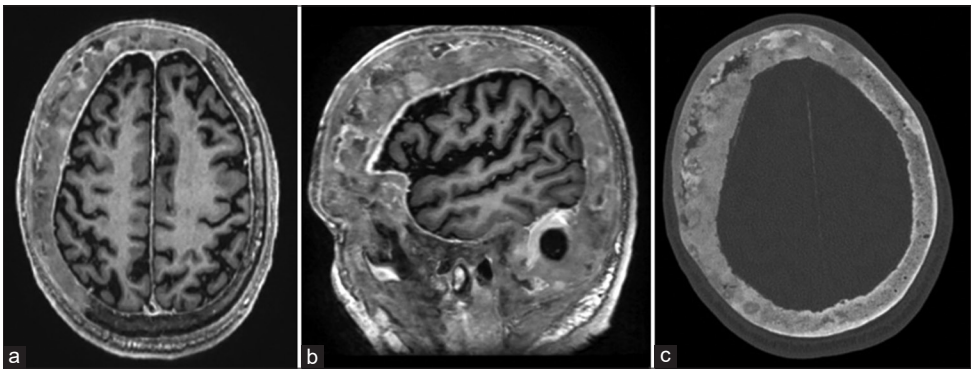

Figure 1: Magnetic resonance imaging T1 with contrast axial (a) and sagittal (b) view showing diffuse calvarial expansion with patchy enhancement in the right frontoparietal, temporal, basilar occipital, basisphenoid, and clival regions. Axial computed tomography bone window (c) showing enlarged diploic space with mixed sclerotic and lytic areas

Access this article online

Received - 21 December 2022
Initial Review - 03 January 2022
Accepted - 02 February 2022

DOI: 10.32677/ijcr.v9i2.3849

Quick Response code

Correspondence to: Anuja Patil, Department of Neurology, Krishna Institute of Medical Sciences, Hyderabad - 500 003, Telangana, India. E-mail: patilanuja1986@gmail.com

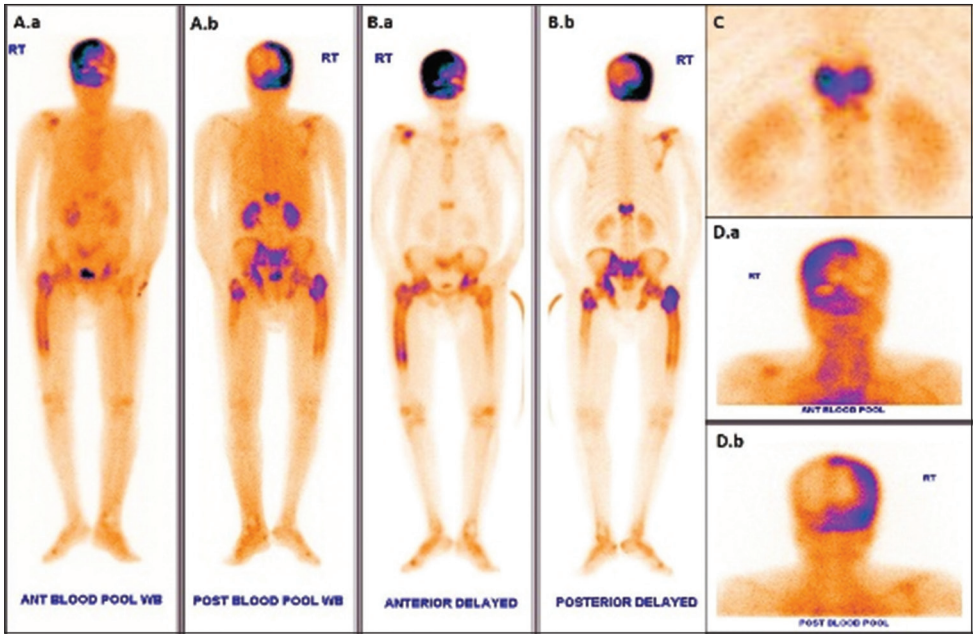

Figure 2: Three phase 99 m Tc bone scan showing increased blood pooling (A.a, A.b) in the right side of calvarium (D.a, D.b), right scapula, D12, sacrum, left hemipelvis, and bilateral proximal femori. Delayed phase (B.a, B.b) showing subsequent diffusely increased tracer uptake. Mouse face appearance of D12 vertebra (c) with increased uptake in body, posterior elements, and spinous process

for 2 months followed by 150 mg monthly. His serum alkaline phosphatase lowered up to 182 units/L at a follow-up of 3 months and had improvement in bulbar symptoms but ptosis and hearing loss persisted.

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Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Vasagiri S, Patil A, Kulkarni P. Paget's disease presenting with multiple cranial neuropathies. Indian J Case Reports. 2023; 9(2):32-33.