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Isolated Temporal Bone's Paget Disease: A Case Report

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Abstract

Paget's bone disease (PBD) is a benign tumor of osteoclasts. We report a case of an isolated temporal bone's Paget disease with unusual evolution to a rapidly functional degradation of vestibulocochlear system. A 60 years old female was admitted for an isolated, left sided mastoid tumefaction without any inflammatory signs. Initial temporal bone computed tomography (CT) showed a left cotton wool appearance without any ossicles lesion or vestibulocochlear extension. Anatomo pathological tests confirmed the diagnosis. The outcome was marked by the occurrence of vestibulocochlear symptoms. Treatment had consisted on subtotal surgery in combination with oral bisphosphonate with improvement of her clinical conditions. The post operative's follow-up was simple without any abnormalities. PBD of temporal bone is a benign disorder. Imaging is considered the most useful diagnosis modality for revealing the disorder that will be confirmed by histological tests. The treatment is based on medical approaches and surgery has few indications.

Keywords: Paget Disease, Bone, Hearing Loss, Temporal Bone Ct, Bisphosphonate

Abbreviations

PBD: Paget's Bone Disease
CT: Computed Tomography
VHIT: Video Head Impulse Test
VOR: Vestibular Ocular Reflex

Introduction

PBD is a disorder of osteoclasts with unknown etiology. It is relatively a common disease affecting up to 4% of individuals over 40 and up to 11% over the age of 80. The bones most frequently affected are the sacrum (57% of cases), followed by the spine (50%), pelvis (43%), and femur (35%) [1]. However, only a small proportion of patients with PDB comes to clinical attention, most likely those with more severe forms [2]. We report through this case the suggestive radiological signs, the histological features, the therapeutic approaches as well as the prognosis of an isolated temporal bone's Paget disease with unusual evolution to a rapidly functional degradation of vestibulocochlear system.

Clinical Case

A 60 years old female without any particular pathological history, presented to our ENT department with one- year history of progressive and painful mastoid tumefaction without any other symptoms such as hearing loss, vertigo, tinnitus and facial paralysis the physical examination had found an isolated, non-tender, left sided mastoid swilling without any inflammatory signs. A complete neurological and ear nose throat examination, otoscopy as well as rhinoscopy, haven't found any abnormalities. A pure-tone audiometry was performed and it was normal. An initial CT scan of the petrous bones demonstrated a cotton wool appearance of left temporal bone which had lost its well-defined contour and the inner and outer tables of the skull appeared indistinct. There was no evidence of ossicles lesions or labyrinth invasion. The right temporal bone was normal (Figure 1 A). A left mastoid biopsy was performed under local anesthesia and the anatomo pathological examination of the specimen confirmed the Paget diagnosis, revealing the typical mosaic pattern with calcified and fibrotic areas. A bone scintigraphy and x-rays were indicated and didn't found any others locations of the disease (Figure 1 B). The patient had a normal level of alkaline phosphatase. We concluded to an isolated left temporal PBD. One month later, the patient presented a hearing loss, vertigo (disappearing progressively and spontaneously after few days, probably due to cerebral compensation system) and tinnitus that prompted an urgent audiogram and ear examination. A video head impulse test (VHIT) was performed hardly because of the pain induced by the rapid movement of the head demonstrating the decreases in vestibular ocular reflex (VOR) gains of the laterals and posteriors semi-circular canals (Figure 2 A). The caloric reflex test had shown a left compensated hyporreflexia measured at 38% (Figure 2 B). Audiometry test revealed a left mixed hearing loss (60 dB) with air bone gap more than 30 dB (Figure 2 C(a)). The patient was operated by the ENT department, a left subtotal mastoidectomy was done under general anesthesia (Figure 3 A) and bone sample was taken for histological analysis and had confirmed the diagnosis (Figure 3 B). The patient was discharged after 4 days. The post operative's follow-up was simple without any abnormalities. After a rheumatology and cardiology consultation, the patient was placed on oral bisphosphonate drugs with improvement and stabilization of her clinical conditions and we recommend to her a left unilateral conventional hearing aid.

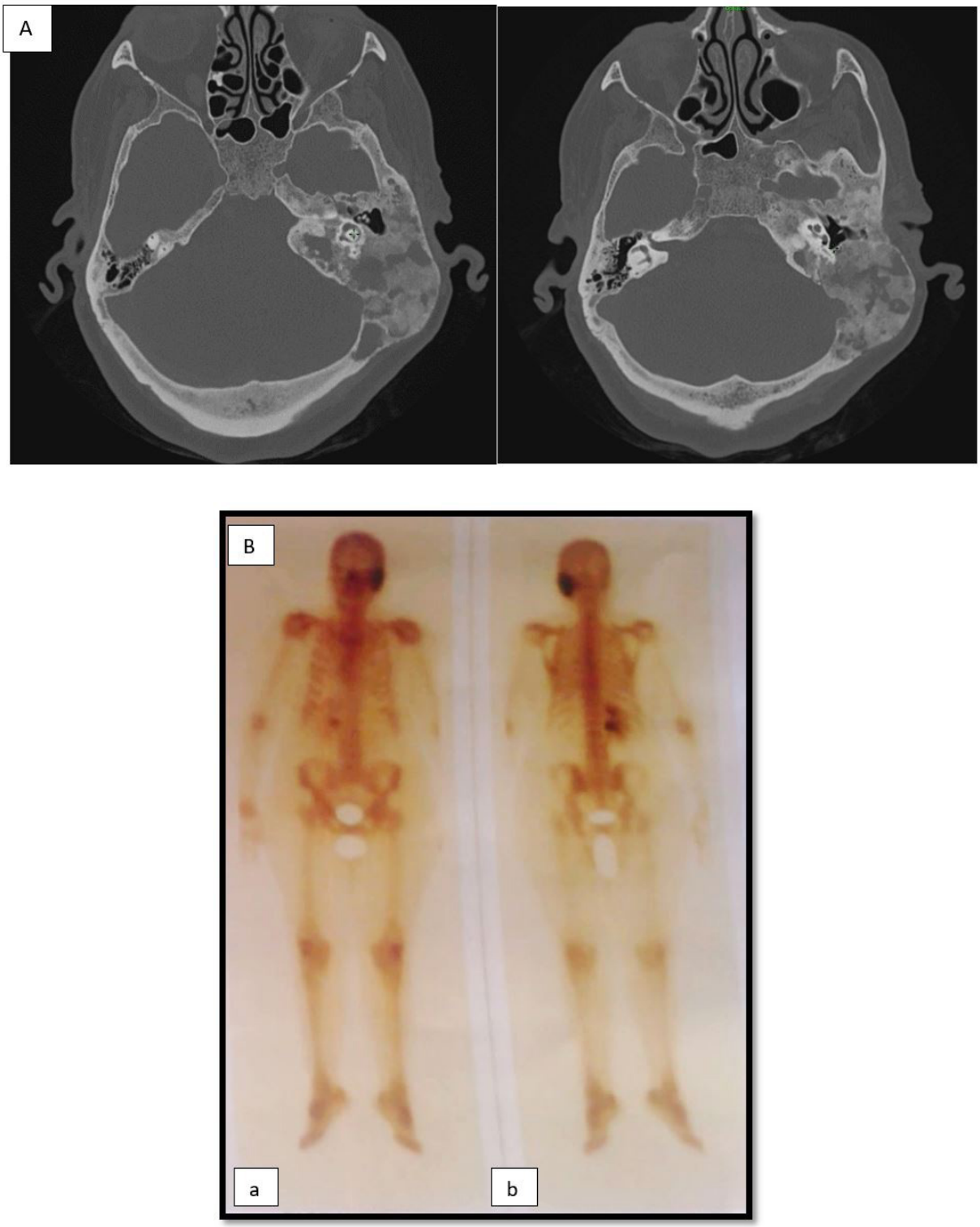
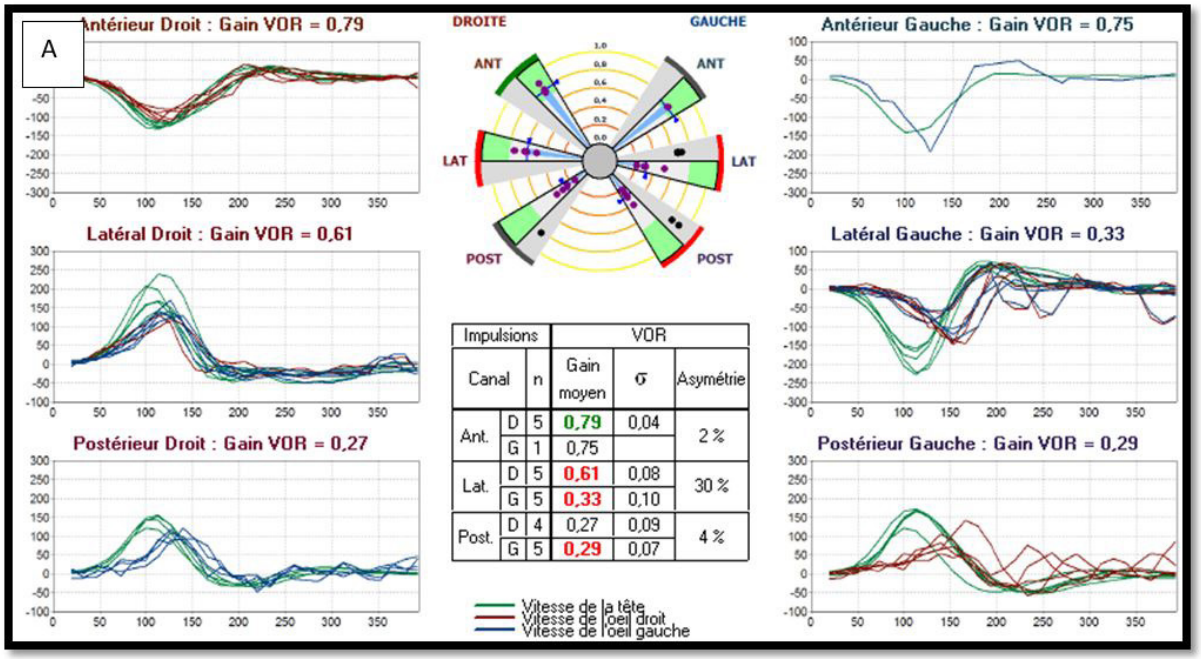
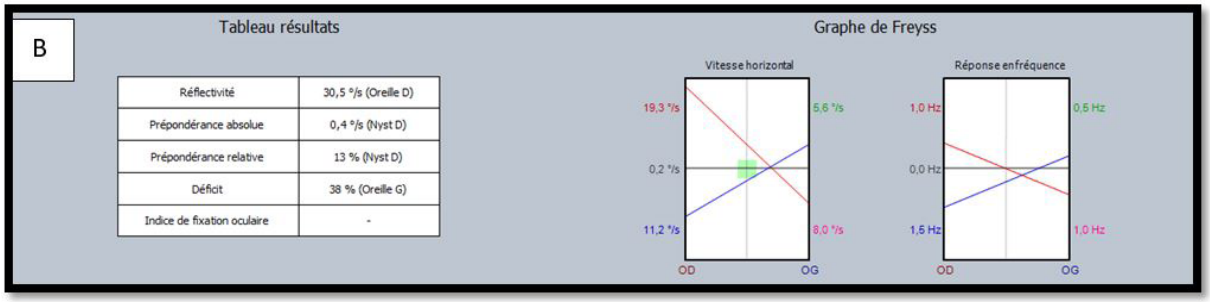


Figure 1: (A) CT scan of petrous bones (axial section): cotton wool appearance of left temporal bone. No evidence of ossicles lesions or labyrinth invasion. The right temporal bone: normal.

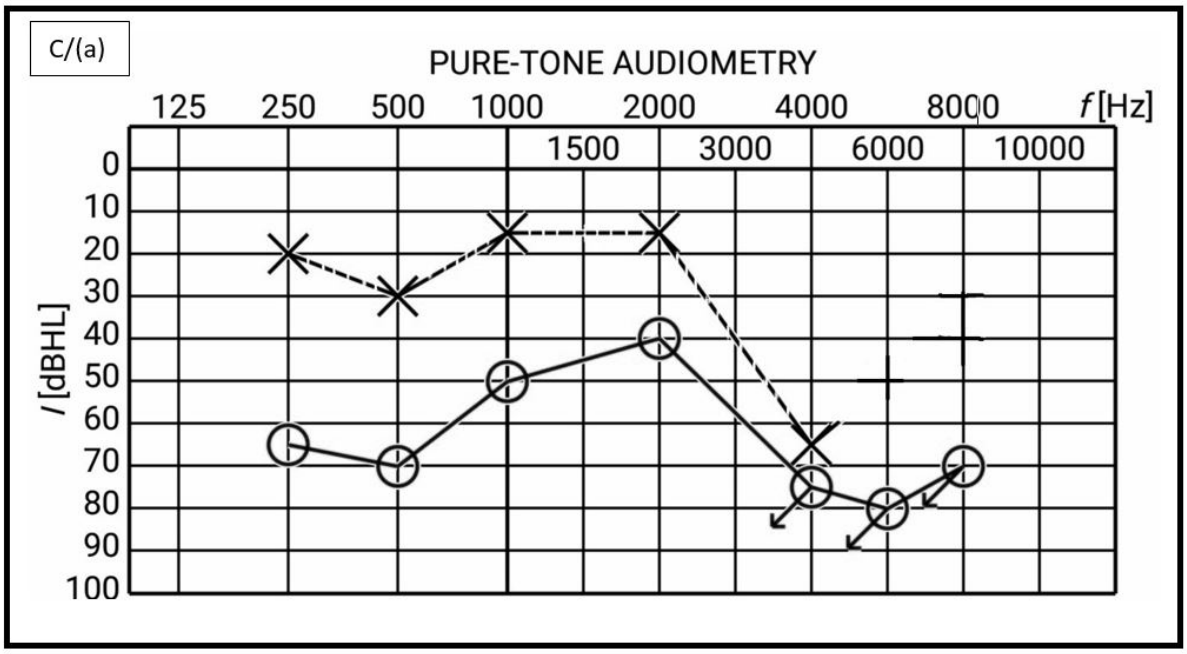
(B) bone scintigraphy: isolated lesion of the left temporal bone a: anterior side of the body b: posterior side of the body



Impulsions			VOR		
Canal		n	Gain moyen	σ	Asymétrie
Ant.	D	5	0,79	0,04	2 %
	G	1	0,75		
Lat.	D	5	0,61	0,08	30 %
	G	5	0,33	0,10	
Post.	D	4	0,27	0,09	4 %
	G	5	0,29	0,07	



(C)



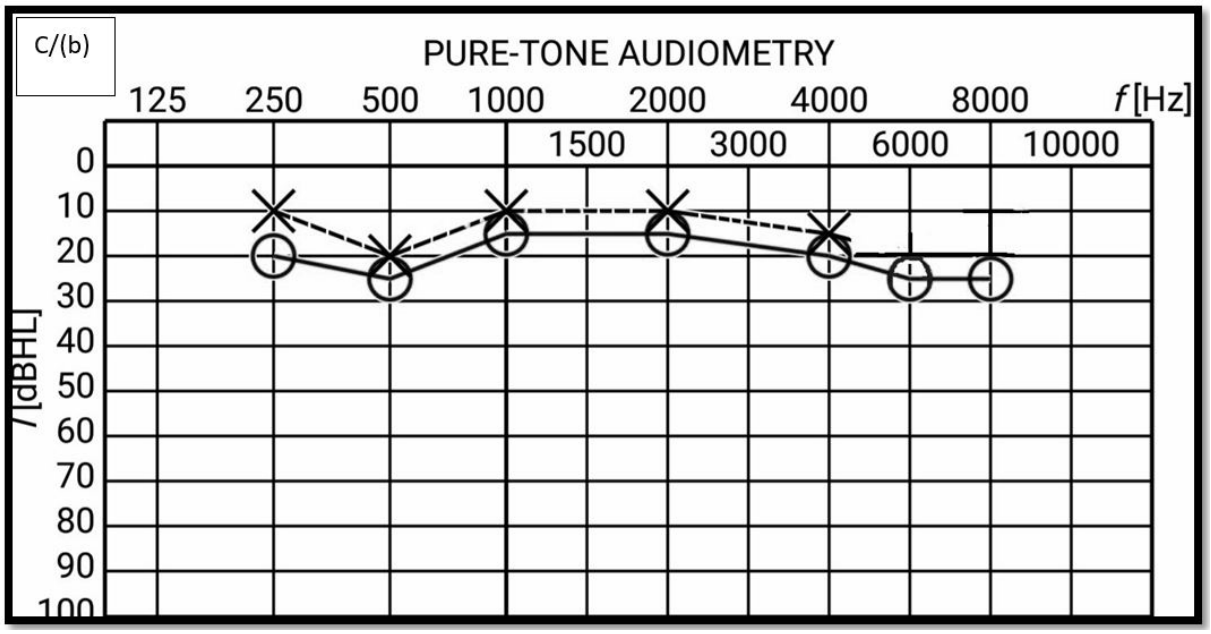


Figure 2: vestibulocochlear explorations

(A) VHIT examination demonstrating the decreases in VOR gains of the lateral and posterior CSC.

(B) caloric reflex test showing a left compensated hyporreflexia measured at 38%.

(C) audiometry test: (a) Left ear: mixed hearing loss with air- bone gap > 30 dB (b) Right ear: normal circles: Air conduction crosses: Bone conduction

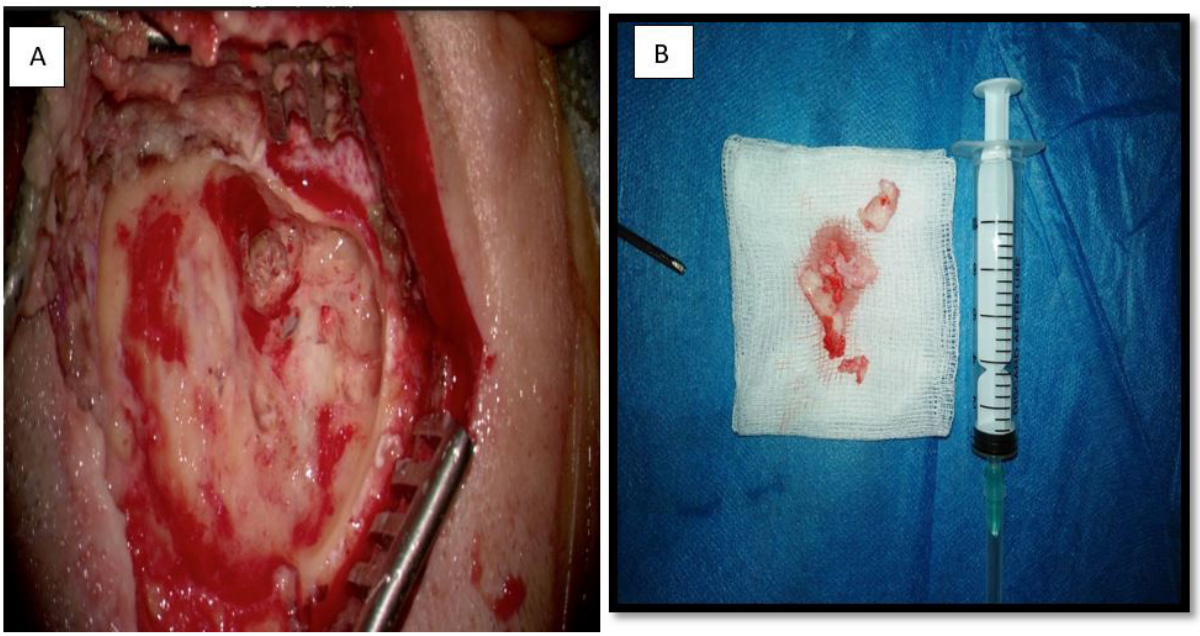


Figure 3: (A) Left subtotal mastoidectomy under general anesthesia (B) Operative specimen

Discussion

PDB is a chronic and focal skeletal disorder, which typically cause enlargement and deformation bones in one or more regions of the skeleton. It is relatively common disease affecting up to 4% of individuals over 40 and up to 11% over the age of 80, males more frequently affected than females. The bones most frequently affected are the sacrum (57% of cases), followed by the spine (50%), pelvis (43%), and femur (35%) [1]. The majority (approximately 3-quarters) of patients are asymptomatic at the time of diagnosis, or presenting localized pain, increased head circumference and complications such as pathologic fractures and neuro otological symptoms [2]. In our case report, our patient developed an isolated temporal bone Paget's disease, not a common site, with a rapidly vestibulocochlear degradation in over one months, she had a left mixed hearing loss and compensated hyporreflexia measured at 38%.

In the literature, the diagnosis of Paget disease of the bone is based essentially on the results of plain radiography, CT scan and anatomo pathological examination. Temporal bone CT scan locate the lesion, reveal its characteristics, the degree of vestibulocochlear extension, and show the differential diagnosis with other otodystrophic lesions of the temporal bone such as otosclerosis and fibrous dysplasia. Progression of the disease into the osteoblastic stage causes cotton-wool spots appearance, similar to the initial CT scan's results found in our case report. Bone scintigraphy should always be included in the investigation of patients with PDB and radiographs of the areas of increased radioisotope uptake should be subsequently made to confirm the diagnosis. PBD bears a striking histologic similarity to otosclerosis, but lesions in end-stage Paget's disease have a classic jigsaw puzzle–like mosaic pattern of "cement lines" [3]. Treatment of PBD does not cure the disease but can provide prolonged periods of remission.

Analgesics, such as nonsteroidal anti-inflammatory medications, is prescribed for pain management. Bisphosphonates, such as alendronate, may be indicated for symptomatic patients to reduce or stabilize bone turnover, promote healing of osteolytic lesions, and improve bone pain. Rarely, osteotomy can be required in patients with focal nerve compression and several conditions in case of other locations than temporal bone's ones, or other focal nerve compression of the spine [4]. In our case report, the subtotal surgical approach was indicated to decompress vestibulocochlear nerve and it was combined with oral bisphosphonate. In general, PBD has a good prognosis but sarcomatous transformation can occur in 5% to 10% of patients with severe polyostotic disease and in 0.15% of patients with more limited involvement. The mean survival time for patients with malignant transformation is less than 1 year [5].

Conclusion

PBD is a benign disorder that can lead to otological symptoms such as impingement of cranial nerves or blockage of the external auditory canal, it can often be a challenge to diagnose because of its insidious presentation. The benign nature of the disease and its location in the skull base should guide the clinician to cautious treatment, reserving surgical intervention either for diagnosis or relief of symptoms. The most dread complication is sarcomatous degenerescence.

Conflict of Interest: None.

Acknowledgments

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