

Case Report

Gradenigo's Syndrome as First Presentation of Solitary Osseous Plasmacytoma of the Petrous Apex

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Abstract

Plasma cell tumors of the skull base are rare in neurosurgical practice. True solitary osseous plasmacytoma of the skull base without development of multiple myeloma is extremely rare. We report a case of typical Gradenigo's syndrome, including left abducens nerve palsy, left facial pain and paresthesia in V₁ and V₂ distribution of trigeminal nerve caused by solitary osseous plasmacytoma of the left petrous apex. The patient was a 46-year-old man who presented with diplopia for two days. Magnetic resonance imaging (MRI) of the brain showed a hyperintense mass on T1-weighted images and slightly hypointense mass on T2-weighted images in the left petrous apex and left parasellar area.

Through a left subtemporal middle fossa approach, subtotal resection of the lesion was performed. Histopathological examination of the lesion revealed plasmacytoma. The patient received 54 Gy radiation for the local tumor. Four months after radiation, the abducens palsy improved. Four years after treatment, the patient remained well with no symptoms or signs of local recurrence or progression to multiple myeloma.

Keywords: Gradenigo's syndrome, petrous apex, plasmacytoma, skull base

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Introduction

Now uncommon complication of acute otitis media is the clinical entity known as Gradenigo's syndrome, first described by Gradenigo in 1904.¹ It consists of a triad including petrous apicitis, ipsilateral abducens nerve palsy, and deep facial pain in the distribution of the trigeminal nerve.¹ As the abducens nerve passes near the apex of the petrous bone, it is in close relation to the trigeminal nerve and both may be implicated by petrositis and adjacent dural inflammation, manifested by facial pain and diplopia. Other lesions located at the petrous-temporal bone apex, especially metastases, fractures, meningiomas, neurofibromas and T cell lymphoma may cause damage or irritate the V₁ division of the trigeminal nerve and the nearby abducens nerve, resulting in Gradenigo's syndrome.¹ Gradenigo's syndrome due to true solitary plasmacytoma of the petrous apex is very rare. We report the first case of typical Gradenigo's syndrome caused by solitary osseous plasmacytoma of the petrous apex.

Case Report

A 46-year-old man presented to our hospital with history of diplopia for two days. He complained of left facial pain and paresthesia in the distribution of the first and second divisions of the left trigeminal nerve for the last two weeks. His neurological examination showed complete left abducens nerve palsy and paresthesia in distribution of V₁ and V₂ divisions of the trigeminal nerve.

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Laboratory tests were normal. Computed tomography (CT) scan of the brain showed destruction of the left petrous apex of the temporal bone (Figure 1). Magnetic resonance imaging (MRI) of the brain showed a hyperintense mass on T1-weighted images and a slightly hypointense mass on T2-weighted images in the left petrous apex and left parasellar area. The mass enhanced after gadolinium administration (Figure 2).

With the initial diagnosis of metastatic lesion or sarcoma, a whole body bone scan was performed and showed increased uptake only in the left petrous apex. Through a left subtemporal middle fossa approach, subtotal resection of the lesion was performed. Histopathological examination of the lesion revealed plasmacytoma (Figure 2). Immunohistochemical staining of neoplastic plasma cells revealed strong and diffuse expression of Ig lambda light chain, but not kappa light chain. The majority of plasma cells expressed CD138 while negative for CD20. Skeletal survey was normal. Bone marrow biopsy was performed. There was no morphological evidence of multiple myeloma.

Immunoelectrophoresis of serum, urinary analysis for Bence-Jones protein, and all immunoglobulins were found to be normal. The postoperative course was uneventful. The patient received 54 Gy radiation for the local tumor. Four months after radiation, the abducens palsy improved.

Immunoelectrophoresis of serum every 6 months and annual bone marrow biopsy have been performed. Four years after treatment, the patient remained well with no symptoms or signs of local recurrence or progression to multiple myeloma.

Discussion

Plasma cell tumors may manifest as one of three pathological entities:^{2,3}

1. Solitary osseous (intramedullary) plasmacytoma (SOP),

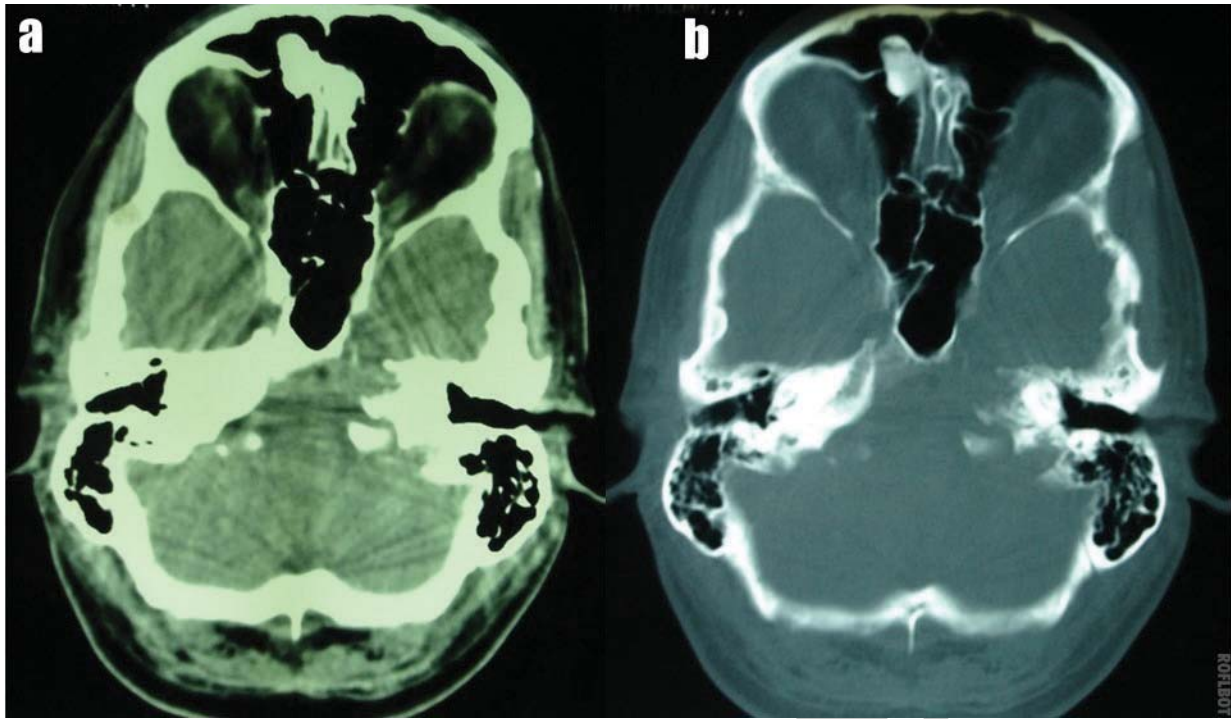


Figure 1. a) Axial non-contrast CT scan of the brain shows destruction of the left petrous apex; b) Axial brain CT with bone window setting shows left petrous apex erosion.

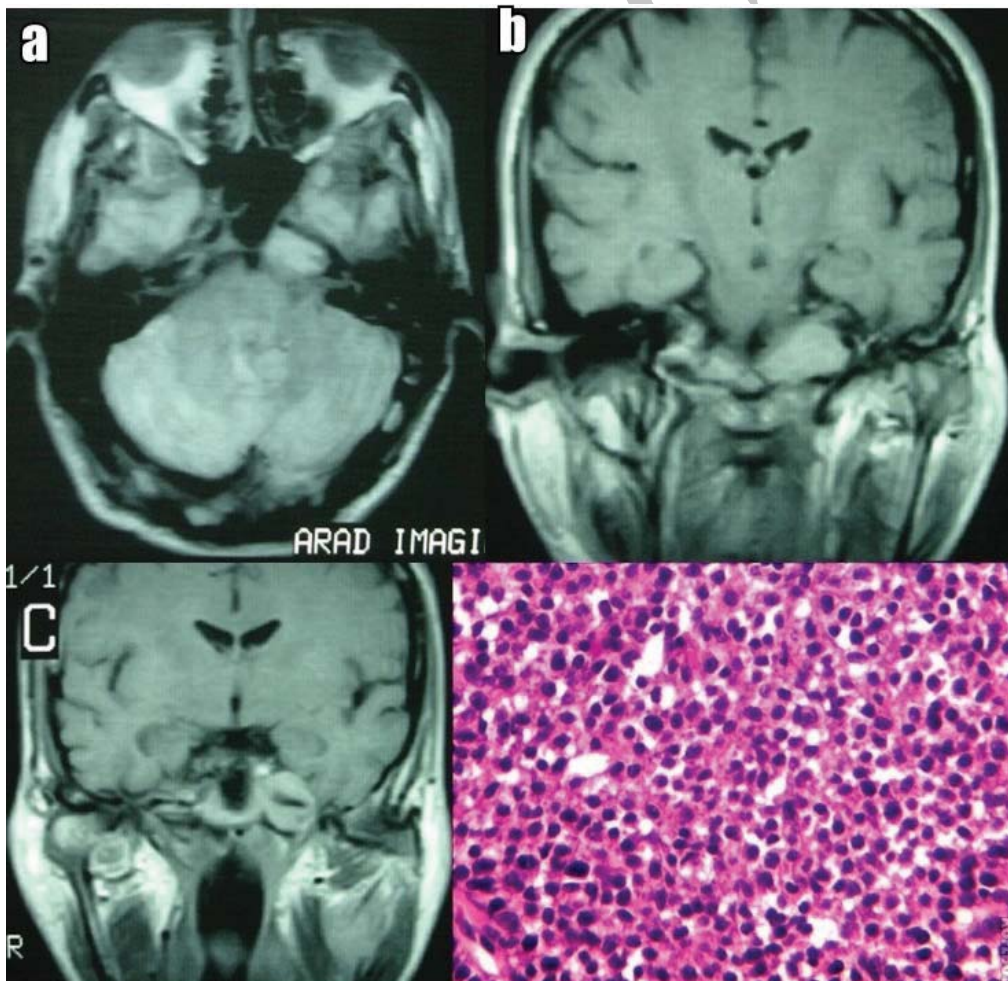


Figure 2. a) Axial T1-weighted MRI of the brain shows a hyperintense mass at the left petrous apex anterior to VII-VIII nerve complex; b) Coronal gadolinium-enhanced MRI of the brain shows an enhancing mass at the left petrous apex; c) Coronal gadolinium-enhanced MRI of the brain shows an enhancing mass at the left parasellar area; d) Histopathology of solitary plasmacytoma (Hematoxylin and Eosin $\times 50$)

2. Solitary extramedullary plasmacytoma (EMP), or
3. Multiple myeloma

Solitary osseous plasmacytoma of the skull base is usually part of a systemic disease.²⁻⁵ It rarely presents as a true localized disease without signs of systemic myelomatosis.²⁻⁵

To our knowledge, this is the first case of Gradenigo's syndrome as a result of true solitary osseous plasmacytoma of the petrous apex. The only other case report of Gradenigo's syndrome due to solitary plasmacytoma has been reported by Bourne, et al.¹ in 1998 in a 45-year-old man with history of multiple myeloma who presented with petrous apex extramedullary plasmacytoma two years after multiple myeloma treatment. Our case was a true solitary osseous plasmacytoma of the left petrous apex without clinical, histologic, or radiological evidence of multiple myeloma either at the time of diagnosis or on further follow-up.

CT scan of the skull base plasmacytoma shows an osteolytic lesion without a sclerotic rim. The tumor is seen as an iso- or hyperdense lesion which demonstrates homogeneous contrast enhancement. On MRI, it is iso-hyperintense on T1-weighted images, iso-hypointense on T2-weighted images and is homogeneously enhancing after administration of gadolinium.^{2,3}

The diagnostic criteria of solitary plasmacytoma are based on a solitary bone lesion on radiography that shows monoclonal plasma cell infiltration on biopsy, and histologically normal bone marrow aspiration. Bone marrow biopsy, skeletal survey and whole body bone scan should be performed to exclude multiple myeloma.^{1,4}

Treatment of solitary plasmacytoma of the skull base is complete removal of the tumor, if possible, followed by radiation therapy.^{3,4} If the tumor is not completely resectable, subtotal resection or biopsy followed by radiotherapy or radiotherapy alone can also control the tumor locally.^{3,4}

Plasmacytoma of the skull base can manifest with a variety of symptoms and signs depending on the site of origin. Gradenigo's syndrome as the first manifestation of skull base plasmacytoma is very rare. We present the first case of Gradenigo's syndrome caused by true solitary osseous plasmacytoma of the petrous apex in a 46-year-old man. We recommend considering solitary osseous plasmacytoma, although unusual, as a potential cause for Gradenigo's syndrome.

References

1. Bourne RRA, Maclaren RE. Intracranial plasmacytoma masquerading as Gradenigo's syndrome. *Br J Ophthalmol*. 1998; **62**: 458 – 459.
2. Pancholi A, Raniga S, Vohra PA, Vaidya V, Prajapati A, Mansingani S. Imaging features of extramedullary plasmacytoma of skull base with multiple myeloma: a rare case. *Ind J Radiol Imag*. 2006; **16**: 29 – 32.
3. Wein RO, Papat SR, Doerr TD, Dutcher PO. Plasma cell tumors of the skull base: four case reports and literature review. *Skull Base*. 2002; **12**: 77 – 85.
4. Bindal AK, Bindal RK, Van Loveren H, Sawaya R. Management of intracranial plasmacytoma. *J Neurosurg*. 1995; **83**: 218 – 221.
5. Ustüner Z, Basaran M, Kiris T, Bilgic B, Sencer S, Sarkar B, et al. Skull base plasmacytoma in patient with light chain myeloma. *Skull Base*. 2003; **13**: 167 – 171.

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