Introduction

A now uncommon complication of acute otitis media is the clinical entity known as Gradengio’s syndrome, first described by Gradengio 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.2 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 V_1 division of the trigeminal nerve and the nearby abducens nerve, resulting in Gradengio’s syndrome.3 Gradengio’s syndrome due to true solitary plasmacytoma of the petrous apex is very rare. We report the first case of typical Gradengio’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 V_1 and V_2 divisions of the trigeminal nerve.

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 postoperative course was uneventful. 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.

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 paracellar area; d) Histopathology of solitary plasmacytoma (Hematoxylin and Eosin × 50)
2. Solitary extramedullary plasmacytoma (EMP), or
3. Multiple myeloma

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

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. 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.

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.

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

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