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Case Report

Acute abducens nerve palsy from a complex retro-odontoid pseudotumor with a protrusion: A case report [☆]

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ABSTRACT

Retro-odontoid pseudotumors represent soft tissue proliferation surrounding the transverse ligament of the atlas, which most commonly results in cervical neck pain or myelopathy due to impingement upon the cervicomedullary junction. The causes of retro-odontoid pseudotumor formation are varied and include metabolic, inflammatory, degenerative, and post-traumatic etiologies. To the best of our knowledge, an abducens nerve palsy as a result of a complex retro-odontoid pseudotumor has never been reported. We discuss a case of a 90-year-old woman who presents with acute lateral gaze palsy with multimodality imaging demonstrating a retro-odontoid pseudotumor with an unusual protrusion which courses superolaterally to the level of the pons and compresses the abducens nerve root entry zone.

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Introduction

Retro-odontoid pseudotumors (RPs) represent non-neoplastic soft tissue proliferation at the atlantoaxial junction surrounding the region of the transverse, apical, and alar ligaments with frequent extension into the retroclival region. RPs have also been described as a periodontoid mass or, in the setting of rheumatoid arthritis, as a pannus. It is thought that the majority of RPs are formed secondary to repetitive injury of

the atlantoaxial junction and its ligaments, which results in chronic inflammation and the formation of fibrotic and granulation tissue [1–3]. The specific diagnoses most commonly attributed to RP formation are rheumatoid arthritis and calcium pyrophosphate deposition disease; however, other etiologies include hemodialysis-associated amyloidosis, pigmented villonodular synovitis, prior fracture, gout, degenerative disc disease, and ossification of the posterior longitudinal ligament [2].

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The clinical sequelae of RPs vary widely. The majority of RPs remain clinically silent; however, as they grow and progressively crowd the cervicomedullary junction, there can be compression of the adjacent spinal cord. In these cases, patients typically experience symptoms including cervical neck pain, paresthesias, and weakness. These symptoms have the potential to be debilitating and cause a poor quality of life. Rarely, compression of the spinal cord could result in death [2,3].

On computed tomography (CT), the typical imaging finding of an RP is a mildly enhancing mass which could cause erosive changes in the adjacent dens. There is a varying degree of calcification. On magnetic resonance imaging (MRI), RPs typically have mild diffuse or rim enhancement, demonstrate T1 prolongation, and have variable T2 signal characteristics [4]. Histologically, RPs demonstrate acellular material resembling degenerated intervertebral discs [5]. Surgical stabilization of the craniovertebral junction has been demonstrated to cause regression of RPs [6].

Here, we report a case of a complex RP with an unusual presentation of acute abducens nerve palsy.

A noncontrast head CT was performed and demonstrated a hypoattenuating focus on the inferolateral aspect of the left hemipons and a partially imaged soft tissue mass with irregular calcification centered at the junction of the clivus and odontoid process. This soft tissue mass partially effaced the ventral cerebrospinal fluid space at the level of the foramen magnum and premedullary cistern (Figs. 1 and 2).

An MRI brain subsequently demonstrated a T2 hypointense mass spanning the retroclival and retro-odontoid regions with a conically shaped, centrally T2 hypointense, peripherally T2 hyperintense protrusion arising from its superior aspect. This protrusion extends through the premedullary cistern and contacts the inferior pontomedullary junction. This contact exerts a mass effect upon the left abducens nerve at its entry zone with gliosis and volume loss at the site of contact (Fig. 3). This mass demonstrates minimal enhancement (Fig. 4).

The patient's left eye was treated with patching. The diplopia has improved but remains unresolved 4 months after presentation. Given her age, she deferred surgical intervention.

Case

A 90-year-old female presented to the emergency department for evaluation of acute diplopia with left lateral gaze palsy. She denied fever, weakness, fatigue, loss of sensation, or facial droop. Her past medical history was remarkable for hyperlipidemia. She was normotensive and afebrile. Physical examination was notable for an inability to abduct the left eye with normal pupillary responses. Her neurological examination was otherwise unremarkable. Routine laboratory testing was also unremarkable.

Discussion

Retro-odontoid pseudotumors (RPs) are soft tissue proliferations at the atlantoaxial junction surrounding the region of the transverse ligament which can be associated with cervical neck pain or compressive myelopathy. Here we show an unusual case of an RP presenting with an acute abducens nerve palsy secondary to a superolateral protrusion which impinged upon the sixth cranial nerve entry zone. The etiology of this protrusion is not entirely clear and may represent cystic degeneration of the RP or an extruded fragment. A superimposed atlantoaxial intradural synovial cyst could be considered in



Fig. 1 – Axial noncontrast head CT demonstrating a hypoattenuating focus within the inferior left hemipons.

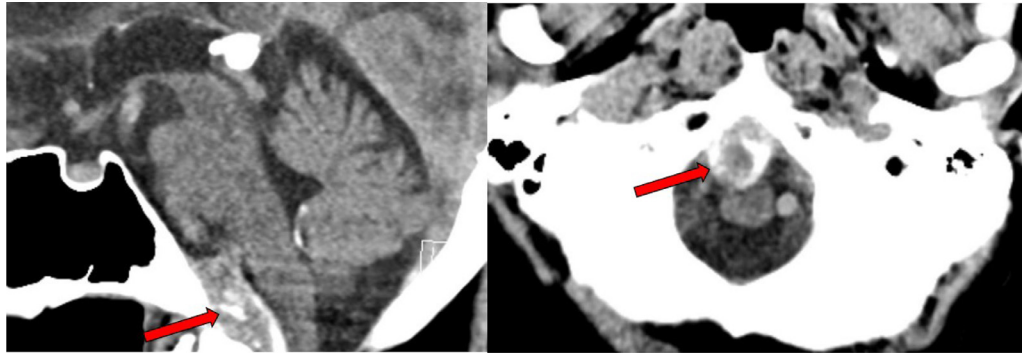


Fig. 2 – Sagittal and axial noncontrast head CTs demonstrating a partially imaged, partially calcified soft tissue mass along the dorsal aspect of the clivus and odontoid process causing partial effacement of the ventral cerebrospinal fluid space at the level of the foramen magnum and premedullary cistern (red arrows).

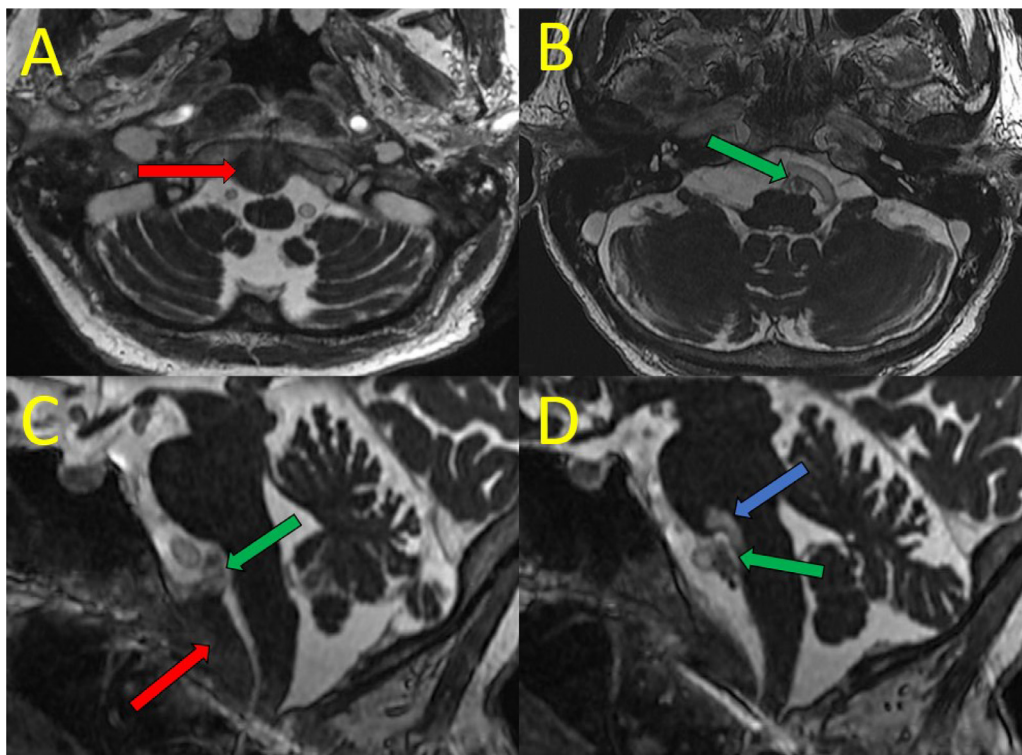


Fig. 3 – 3D axial (A, B) and sagittal (C, D) Fast imaging employing steady-state acquisition (FIESTA) MRI of the brain at the level of the posterior fossa. These images demonstrate a T2 hypointense mass behind the clivus which corresponds to the mass on CT (red arrow, A and C). There is a conically shaped protrusion arising from the superior aspect of this mass which demonstrates a T2 hypointense core and a T2 hyperintense periphery (green arrow, B, C, and D). The protrusion courses to the left of midline (image D) where it contacts the inferior pontomedullary junction with associated gliosis and volume loss at the site of contact (blue arrow). There is a mass effect and slight distortion of the left abducens nerve at its entry zone at the site of contact (image B).

the differential diagnosis. However, these typically arise from the synovium of the atlantoaxial joint which is relatively distant [7]. Additionally, this protrusion appears to arise directly from the RP. As such, the presence of an atlantoaxial intradural synovial cyst is felt to be less likely.

A systematic review of RPs studying 36 articles involving a total of 62 patients did not include a single case of lateral gaze palsy. Instead, these cases most frequently describe cervical neck pain and symptoms of myelopathy [8]. This case's unusual presentation serves as a demonstration of the vary-

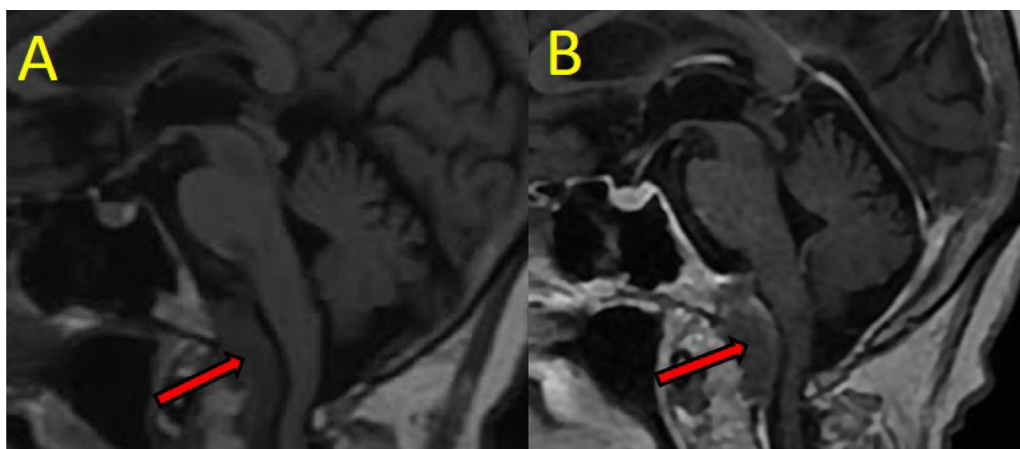


Fig 4. – Axial T1 MRI of the brain without (A) and with (B) intravenous contrast demonstrates minimal postcontrast enhancement of the retroclival mass.

ing effects that an RP can have by way of local mass effect. In conclusion, in patients presenting with a lower cranial nerve palsy, especially if they also have the more classic symptoms of cervical neck pain and myelopathy, an RP should be considered within the differential diagnosis.

Patient consent

Informed consent was obtained directly from the patient for publication of their case.

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