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Surgical Neurology International

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Case Report

A case of nonrheumatoid retro-odontoid pseudotumor in Klippel-Feil syndrome with C1 occipitalization

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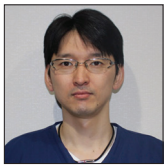

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Received : 25 September 2022
Accepted : 06 October 2022
Published : 21 October 2022

DOI
10.25259/SNI_891_2022

Quick Response Code:

ABSTRACT

Background: Patients with both nonrheumatoid retro-odontoid pseudotumors (ROPTs) and congenital craniocervical junction (CCJ) abnormalities are rare. Here, a 73-year-old female presented with neck pain and myelopathy due to MR-documented ROPT with intramedullary hyperintensity at the CCJ warranting an occipital-cervical fusion.

Case Description: A 73-year-old female originally developed occipitalgia and became quadriparetic within the subsequent 7 months. The cervical MR showed a ROPT with intramedullary hyperintensity at the CCJ. Further, the CT demonstrated C1 occipitalization and a congenital C2-3 fusion without radiological instability. After she underwent an occipito-C2 fusion, her symptoms improved.

Conclusion: For patients with C1 occipitalization and a Klippel-Feil syndrome, ROPT may occur due to loading of C1-2 complex. These patients typically favorably respond to occipito-C2 fusion.

Keywords: C1 assimilation, Craniocervical junction, Klippel-Feil syndrome, Nonrheumatoid retro-odontoid pseudotumor

INTRODUCTION

Nonrheumatoid retro-odontoid pseudotumors (ROPTs) with/without instability are common in elderly population.[6,9] They may also be associated with diffuse idiopathic skeletal hyperostosis or ankylosing spinal hyperostosis in the middle and lower cervical spine.[6,8] Here, we encountered a 73-year-old female with a rare craniocervical junction (CCJ) malformation consisting of C1 assimilation and a C2–3 Klippel-Feil (K-F) syndrome along with ROPT who required an occipito-C2 fusion.

CASE REPORT

A 73-year-old female presented with a 7-month history of occipitalgia and a progressive quadriparesis. The initial neurological examination showed severe myelopathy. The cervical MR showed retro-odontoid mass with an intramedullary T2 high cord signal [Figure 1a]. The 3D-CT at the CCJ showed C1 assimilation and congenital Klippel-Feil fusion of the C2–3 vertebrae [Figure 1b]. Dynamic X-rays showed no frank instability, but the atlantodental interval

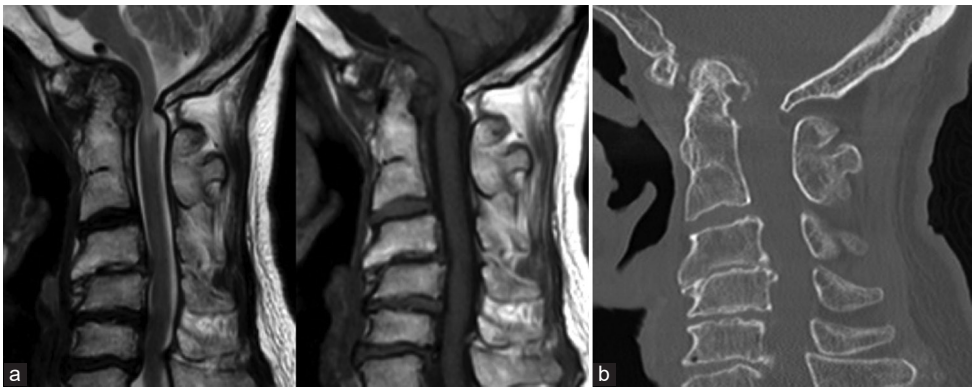

Figure 1: (a) MRI showed retro-odontoid mass with intramedullary T2-weighted image high signal. (b) Three-dimensional computed tomography (3D-CT) findings at craniocervical junction showed C1 assimilation and C2-3 fused vertebrae.

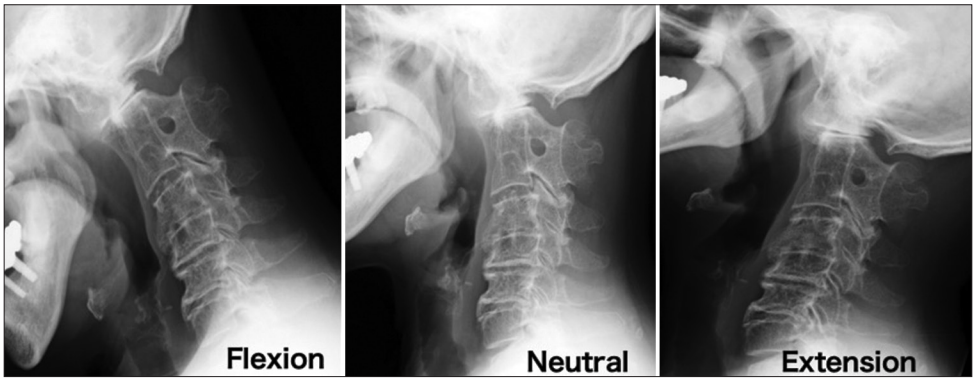

Figure 2: Dynamic X-ray showed no obvious radiological instability with an atlantodental interval of 3.5 mm, but an instability angulation with a range of motion of 20°.

(ADI) was 3.5 mm, with 20° of angulation [Figure 2]. She underwent decompressive surgery of the foramen magnum including a C1 laminectomy followed by occipitocervical fusion with intraoperative neurophysiological monitoring [Figure 3a]. Postoperatively, her symptoms improved. Three months postoperatively, the MRI showed a reduction in ROPT and slightly resolution of the high cord signal [Figure 3b].

DISCUSSION

In patients with C2–3 fusion, especially with additional C1 occipitalization, several secondary anomalies include atlantoaxial dislocation, basilar invagination, Chiari malformation, and syringomyelia can be identified.[3,7] Such cases are usually young and present with neurological symptoms, but this case is interesting because it was an elderly person who presented with ROPT. ROPT has been reported with or without atlantoaxial instability[1,2,4] [Table 1]. Some reports advocate a combination of indirect decompression and immobilization in similar cases.[10]

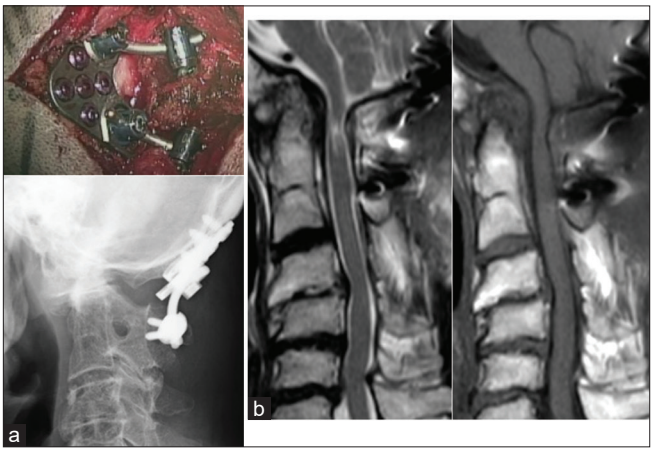

Figure 3: (a) Intraoperative findings and postoperative X-ray show occiput-cervical fixation. (b) MRI after three months surgery showed a reduction in ROPT.

Reports of ROPT have been increasing in recent years and decompression alone or combination with immobilization is controversial for its treatment.[6] ADI is used for the

Table 1: Report of the ROPT in Klippel-Feil syndrome with C1 occipitalization.

Author	Age	Sex	Surgery	ROPT reduction
Buttiens *et al*.,[1] 2018	67	Female	C1-C fusion	Yes
Gandbhir *et al*.,[2] 2021	NA	NA	O-C fusion	Yes
Lagares *et al*.,[4] 2005	69	Male	O-C fusion	Yes
Present case	73	Female	O-C fusion	Yes

NA: Not available, ROPT: Retro-odontoid pseudotumors

definition of instability of C1–2 and there is no definition of instability of angulation. The development of ROPT involves the degenerative process and adjacent segment disease of the ventral ligaments of the CCJ.[5] In this case, the presence of angulation instability of C1–2 was demonstrated.

CONCLUSION

A 73-year-old female with myelopathy attributed to a retro-odontoid pseudotumor with cord compression (i.e., high cord signal) and a congenital C2–3 fusion with C1 assimilation was successfully treated with foramen magnum decompression and occipito-C2 fusion.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Sakamoto K, Uchikado H, Hanada H, Inoue R, Nii K, Higashi T. A case of nonrheumatoid retro-odontoid pseudotumor in Klippel-Feil syndrome with C1 occipitalization. Surg Neurol Int 2022;13:480.