CASE REPORT

Jolt accentuation of neck pain: a novel physical finding for crowned dens syndrome

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SUMMARY

Crowned dens syndrome (CDS), a pseudogout attack involved with atlantoaxial joint, mimics meningitis, because jolt accentuation of headache, a physical sign for meningitis, is frequently considered mistakenly as 'positive' in CDS patients. Our patient with CDS experienced multiple ambulance transports and underwent lumbar puncture for suspected meningitis because of positive result of jolt accentuation of headache. We found that the patient actually had jolt accentuation of neck pain from CDS and treated her successfully. The characteristic physical finding produced by axial neck rotation in CDS patients is not headache, but a jolt accentuation of neck pain.

BACKGROUND

Crowned dens syndrome (CDS) is acute neck pain caused by pseudogout of the atlantoaxial junction in cervical spines and requires only conservative a short-term management with non-steroidal anti-inflammatory drugs or steroids.¹ However, because its symptoms mimic those of several diseases, including meningitis,^{2–4} CDS patients often receive

unnecessary tests, including invasive diagnostic procedures.

We report a case of CDS who had been brought through ambulances four times between several hospitals and received extensive tests, including lumbar puncture, until the correct diagnosis was made. Here, we propose a novel physical finding for making the correct diagnosis in patients with CDS.

CASE PRESENTATION

A woman aged 88 years with hypertension presented with a 4-day history of severe neck pain to the first hospital by ambulance. In the first hospital, she underwent extensive tests. There was no evidence of acute coronary syndrome and cerebrovascular disorder, and thus, she was brought through first emergency transport to another community hospital.

In this second hospital, meningitis was suspected because jolt accentuation was positive on physical examination. Thereafter, she was brought through third transport to another general hospital for further evaluation of possible meningitis. However,

Figure 1 (A) Sagittal CT scan image of the neck showing linear calcification (red arrows). (B) Axial CT scan image showing curvilinear calcifications of the transverse ligament of the atlantoaxial joint (red arrow). (C) Coronal CT scan image showing crown-like calcifications around the odontoid process (red arrow).

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cerebrospinal fluid test was performed, showing normal results. Since the cause for the neck pain was unclear, she was brought through fourth transport to our hospital.

On examination, she appeared ill and febrile up to 37.5°C. There was the limitation of passive neck rotation to all directions. Jolt accentuation of the increased neck pain by axial rotations of the neck was observed. Peripheral white cell count was 8880/μL and C reactive protein was 19.16 mg/dL. Cervical spine CT showed calcification along the transverse ligament of the atlantoaxial joint, indicating CDS (figure 1A–C).

OUTCOME AND FOLLOW-UP

She was admitted to our hospital with the treatment of oral loxoprofen. On day 10, the neck pain completely disappeared and she was discharged.

DISCUSSION

Our case illustrates the importance of making a diagnosis of CDS swiftly since it mimics many important diseases such as meningitis to avoid extensive diagnostic tests. CDS was reported as acute neck pain from atlantoaxial joint inflammation due to hydroxyapatite or calcium pyrophosphate dihydrate crystal deposition.¹ CDS is increasingly recognised as an important cause for acute neck pain with fever among elderly people.

Many patients with CDS undergo lumbar puncture since meningitis is suspected because of 'positive' jolt accentuation of headache. However, this interpretation in CDS patients leads to misdiagnosis as meningitis.^{2–4} In our case, lumbar puncture was performed because 'jolt accentuation of headache' was considered as positive.

Jolt accentuation of headache which consists of worsening of headache by shaking the neck at a frequency of two to three times per second is frequently used for diagnosis of meningitis.⁵ It had a sensitivity of only 21% and a specificity of 83% for the diagnosis in recent studies.⁶ It can be falsely positive if CDS would be asked whether the neck pain would become worse when the neck rotation is provided as it happened in our case.

Therefore, the new physical term, jolt accentuation of neck pain, should be used for correctly differentiating CDS from meningitis. Patients with CDS should have increased neck pain by letting patients rotate the neck. Jolt accentuation of neck pain is different from headache by axial rotation because pain region was confined to the neck.

In conclusion, it is important to consider CDS in the differential diagnosis of acute neck pain to avoid inappropriate emergency transports and unnecessary tests, including lumbar puncture. Therefore, better understanding of physical finding

for CDS is required and jolt accentuation of neck pain may be helpful for the differential diagnosis of neck pain.

Patient's perspective

I had severe neck pain and it was getting worse by the neck motion. Several doctors thought that it might be meningitis and thus I was transported by ambulances multiple times. But I thought it might be neck illness. Finally, I had got a correct diagnosis by a doctor who examined me very carefully and warmly.

Learning points

- Crowned dens syndrome (CDS) should be positively considered in patients who have acute neck pain to avoid inappropriate emergency transports and unnecessary tests.
- If CDS is diagnosed, the most of its patients are treated by only non-steroidal anti-inflammatory drugs.
- Jolt accentuation of neck pain may be helpful for the differential diagnosis of neck pain.

Contributors MK cared the patient and MK and YT wrote the manuscript.

Competing interests None declared.

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