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QUIZ CASES

A rare cause of Horner's syndrome: cervicothoracic spinal root cysts

Dilek Top Karti[1] · Omer Karti[2] · Nese Celebisoy[3]

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Abstract

Objection We herein report cervicothoracic spinal root cysts as a rare cause of Horner's syndrome.

Case report A 62-year-old woman was admitted to our neuro-ophthalmology clinic complaining of drooping of her right upper eyelid. The history, physical examination, and laboratory tests were normal. The extraocular movements were full. The right eyelid was ptotic and the right pupil was smaller than the left. Right Horner's syndrome was diagnosed by the neuro-ophthalmologic examination and pharmacological tests. Cervical magnetic resonance imaging showed multiple spinal nerve root cysts between C7 and T1 segments of the spinal cord.

Conclusion This report showed for the first time that cervicothoracic spinal root cysts could cause Horner's syndrome and should be kept in mind in performing neuroimaging studies.

Keywords Horner's syndrome · Oculosympathetic pathway · Ptosis · Spinal root cyst · Tarlov cyst

Introduction

Horner's syndrome was first noted experimentally by Claude Bernard in 1852. Its classical triad consists of partial ptosis and miosis, but normally reactive pupil and facial anhidrosis on the affected side [1]. This syndrome may occur as a result of any interruption in the oculosympathetic pathways that extends from the brain to the eye [1, 2]. The purpose of this report is to present an unusual case with Horner's syndrome due to spinal root cysts.

Case report

A 62-year-old woman was admitted to our neuro-ophthalmology clinic complaining of partial right ptosis. Her complaint commenced 2 months ago and her past medical history was unremarkable. On admission, general physical examination was normal. On neuro-ophthalmologic examination, best corrected visual acuity with the Snellen chart was 10/10 in both eyes with correction. Both slit-lamp and fundus examinations gave normal results. There was right-sided ptosis. Eye movements were within normal limits. There was anisocoria which was more prominent in the darkness. The pupil diameter in dim light in a darkened room was approximately 2 mm in OD and 3.5 mm in OS. The margin reflex distance 1 (MRD 1) was 2 mm in OD and 4 mm in OS (Fig. 1a). The light and near reflexes were present on either side. Pharmacological test with topical 0.5% apraclonidine (Iopidine, Alcon, Fort Worth, TX, USA) was performed for the diagnosis of Horner's syndrome. After topical application, the right pupil dilated from 2 to 5 mm in dim light in a darkened room. Besides pupillary dilatation, mild ptosis of the right eyelid improved. The right MRD 1 increased from 2 to 4 mm (Fig. 1b). Horner's syndrome was diagnosed and laboratory investigations were initiated. Blood tests including biochemistry and blood count were within normal limits. Comprehensive radiological imaging, including chest X-ray, carotid Doppler ultrasonography, thorax and brain computed tomography (CT), brain magnetic resonance imaging (MRI), and magnetic resonance angiography of the head and neck, gave normal results. However, cervical MRI revealed multiple spinal root cysts between C7 and T1 segments which were thought to be the cause of right-sided Horner's syndrome

✉ Omer Karti
kartiomer@gmail.com

[1] Department of Neurology, Bozyaka Training and Research Hospital, İzmir, Turkey

[2] Department of Ophthalmology, Bozyaka Training and Research Hospital, Saim Çıkrıkcı Cad. No. 59, Bozyaka, İzmir, Turkey

[3] Department of Neurology, Ege University, İzmir, Turkey

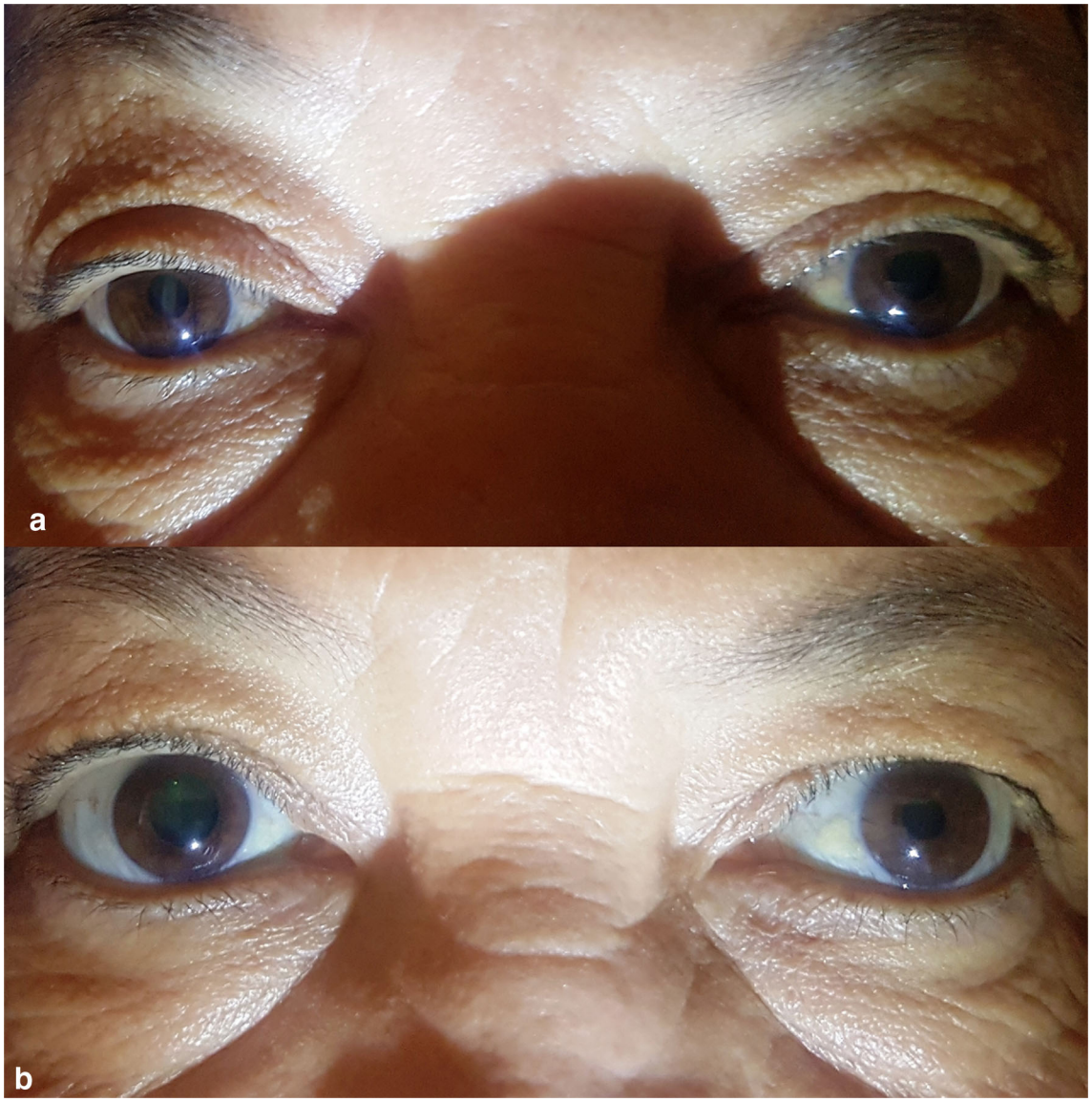

Fig. 1 Clinical photographs obtained in dim light in a darkened room demonstrating right Horner's syndrome. Mild ptosis and miosis in the right eye before topical application of 0.5% apraclonidine (**a**). Pupillary dilatation and improvement in ptosis in the right eye after topical application of 0.5% apraclonidine (**b**)

(Fig. 2). Examination by an experienced spinal surgeon confirmed the Tarlov cysts. As she did not have any complaints except mild ptosis of the right upper eyelid and neurological examination was normal, medical or surgical treatment was not recommended. The patient was monitored periodically. She remained neurologically asymptomatic. An additional complaint or symptom was not observed throughout the follow-up period.

Discussion

Spinal nerve root cysts, also known as "Tarlov cysts," which were first described by Isadore Tarlov as an incidental finding during his autopsy study in 1938, are cerebrospinal fluid (CFS) filled cysts of the spinal cord [3]. The prevalence is 4–9% in the adult age group [4]. Tarlov cysts are more often found in the sacral roots of the spinal cord [5] but are also rarely reported in cervical, thoracic, and lumbar roots [6–9]. Though mostly asymptomatic and coincidentally found on magnetic resonance imaging (MRI), they may become symptomatic when the nerve structures within the cyst or surrounding tissue are mechanically irritated. Symptomatic cases are very rare and constitute 1% of cases [6]. The cyst-related symptoms may vary depending on the location. Cases with sacral cysts usually suffer from low back or sacral pain, coccydynia, sciatica, and cauda equina syndrome. On the other hand, cases with cervical and thoracic cysts may suffer from

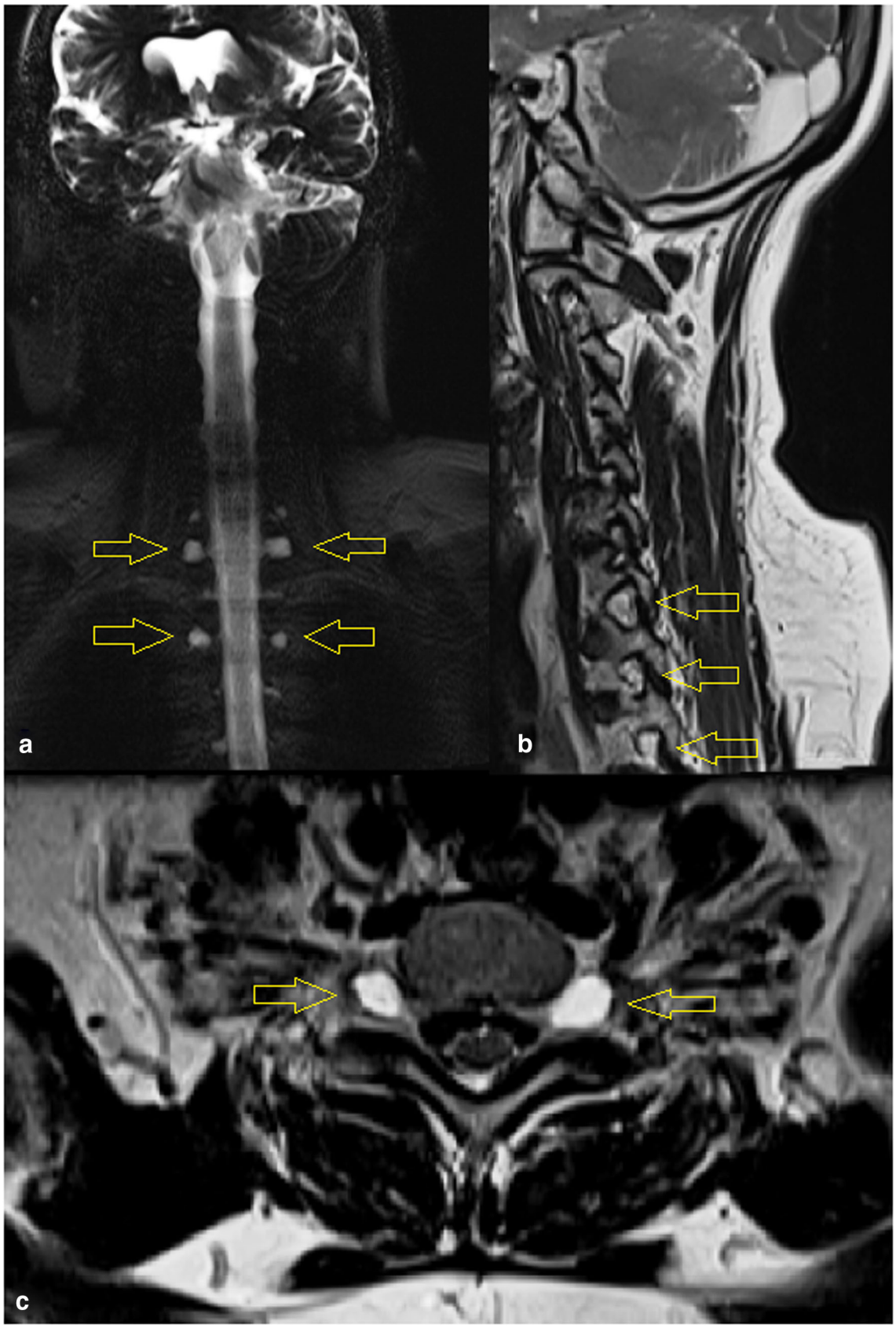

Fig. 2 Magnetic resonance imaging showing multiple (C7–T1) spinal root cysts (**a**). T2-weighted sagittal (**b**) and axial (**c**) images depicting hyperintense lesions (yellow arrows point to the lesions)

cubital tunnel syndrome, cervicobrachialgia, and thoracic outlet syndrome [5–10].

Several hypotheses such as trauma, arachnoidal proliferation, inflammation, and developmental or congenital disorders have been proposed for the etiology of cyst formation, but the real etiology remains unclear [11–16]. Histological studies have revealed that the cyst's wall contains neural elements, such as peripheral nerve fibers or ganglionic cells covered with meningeal epithelium. These studies have also stated that space within the cyst may have potential communication with the subarachnoid space [17].

MRI is the currently preferred method for the diagnosis of the cysts. The classical MRI findings of spinal root cysts consist of low signal on T1-weighted images or high signal on T2-weighted images. In addition, MRI and CT without contrast are performed for differential diagnosis for detecting extradural spinal masses [18].

Periodical follow-up is usually sufficient in asymptomatic cases in general practice. However, both surgical and conservative management techniques (oral and epidural steroid injection) have been proposed in symptomatic cases [18, 19].

As far as we know, our patient is the first in the literature with Horner's syndrome due to cervicothoracic spinal root cysts. The second neuron of the sympathetic pathway emerges from the spinal cord through the C8–T1 anterior roots to reach the superior cervical ganglion. The cysts filled with CSF might lead to the development of Horner's syndrome through stretching or compressing the sympathetic fibers within the spinal roots. The difficulty for this patient is the presence of bilateral cysts with one-sided findings. As no other causal pathology could be revealed, Horner's syndrome was ascribed to the cysts involving C7–T1 segments.

In conclusion, cervicothoracic spinal root cysts must be kept in mind in patients with Horner's syndrome with no other casual pathology.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed consent The authors obtained consent from the patients for publishing the photo.

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