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An Unusual Presenting Symptom in an Atypical Case of Acute Inflamatory Polyradiculopathy: Facial Pruritus

Atipik Akut Enflamatuvar Poliradikülopati Olgusunda Olağandışı Bir Semptom: Fasiyal Kaşıntı

Ayşegül Gündüz, Gülsün Ersen, Başak Yılmaz, Feray Karaali-Savrun

İstanbul University Cerrahpaşa Faculty of Medicine, Department of Neurology, İstanbul, Turkey

Summary

Itchiness is the desire for mechanical irritation of the skin and it may result from various dermatological and sometimes neurological diseases. Neurological causes for itch are relatively unknown and frequently overlooked. Here, we report a case of possible acute inflamatory polyradiculopathy who had facial pruritus and diplopia as the initial symptoms. Diagnosis of our patient was delayed because initial diplopia and pruritus complaints were considered as nonspecific and unrelated to the nervous system. We suggest that pruritus may be a remarkable neurological finding especially when it is accompanied by other neurological symptoms. (Turkish Journal of Neurology 2015; 21:74-6)

Key Words: Acute inflamatory polyradiculopathy, pruritus, itching, diplopia

Özet

Kaşıntı, çeşitli nörolojik ve dermatolojik hastalıklar sonrasında gelişen, deride mekanik irritasyon için duyulan istektir. Kaşıntının nörolojik nedenleri görece az bilinir ve çoğunlukla gözden kaçırılır. Burada, başlangıç semptomu olarak fasiyal kaşıntı ve çift görme olan olası akut enflamatuvar poliradikülopati olgusu sunulacaktır. Başlangıç semptomu olan fasiyal kaşıntı ve çift görme özgün ve sinir sistemiyle bağlantılı olmadığı düşünüldüğünden hastamızın tanısı gecikmiştir. Kaşıntının, özellikle diğer nörolojik belirtiler eşlik ettiğinde dikkate değer bir nörolojik bulgu olabileceğini ileri sürmekteyiz. (Türk Nöroloji Dergisi 2015; 21:74-6)

Anahtar Kelimeler: Akut enflamatuvar poliradikülopati, pruritus, kaşıntı, diplopi

Introduction

Itchiness is the desire for mechanical irritation of skin and it may result from various dermatological and sometimes neurological diseases. Neurological causes for itchiness is relatively unknown and frequently overlooked. Neuroanatomy of itchiness is anticipated to be similar to that of pain (1,2). Therefore, it would not be surprising that lesions involving peripheral or central somatosensory system may also cause neuropathic itch.

Here, we report a challenging case of possible acute inflamatory polyradiculopathy who had facial pruritus and diplopia as the initial symptoms and aim to discuss the possible underlying mechanisms.

Case Report

A 29 year-old male patient was admitted with double vision, ptosis, pruritus on the left side of face and forehead; fatigue, difficulty with sitting up and urgency. His first complaints were double vision and pruritus on the left side of face, which developed one month before the admission. He claimed that diplopia occurred when he looked to the right side. His complaints progressed within a week, causing lateral and upward deviation of the left eye during neutral position. Pruritus got worse in severity and expanded to involve forehead and left side of neck. Ptosis, weakness which caused difficulty in sitting up or standing up, shoulder and arm pain and urinary urgency were additional complaints which developed within the third week of disease. His

Address for Correspondence/Yazışma Adresi: Ayşegül Gündüz MD, İstanbul University Cerrahpaşa Faculty of Medicine, Department of Neurology, İstanbul, Turkey
Phone: +90 212 414 31 65 E-mail: draysegulgunduz@yahoo.com
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personal or family history did not reveal remarkable diseases. He had common cold symptoms 10 days before the development of other symptoms and they improved without treatment. He did not have a history of flu, diarrhea or toxin exposure.

Neurological examination on the day of admission showed lateral deviation of the left eye, right-sided ptosis, proximal extremity and axial muscle weakness with absent bilateral patella and Achilles reflexes. Superficial touch and pain sensation were normal whereas vibration sense over lower extremities got prolonged and position sense was abnormal. He did not have pyramidal findings or dysmetria. He was unable to walk because of proximal weakness and thus needed assistance.

We performed electrophysiological and radiological investigations to further localize the lesion. Routine peripheral nerve motor and sensory conduction studies, conduction studies over axilla and Erb segments, low frequency repetitive stimulation studies of accessory and facial nerves and high frequency stimulation of ulnar nerve were within normal limits. Regarding nerve conduction studies, the only abnormal finding was slightly prolonged F waves recorded over the tibial nerve. F waves of median and ulnar nerves were within the normal values and the persistencies of F waves were not reduced. Needle electromyography showed normal motor units with weak interference pattern over axial and proximal extremity muscles. Latency and amplitude values of median somatosensorial evoked potentials (SEP) were normal whereas tibial SEP showed bilateral prolonged latencies with normal amplitudes. Although cranial and cervical magnetic resonance imaging (MRI) were also normal without any contrast enhancement, lumbar MRI demonstrated weak contrast enhancement of some of the lumbosacral radices (Figure 1). As electrophysiological and radiological findings supported that origin of findings was probably proximal parts of the nerves or radices, we proceeded with the investigations regarding possible etiological factors. Differential diagnosis included infections like diphteria, human immunodeficiency virus (HIV), lepra, inflamatory or toxic polyneuropathies, connective tissue disorders, sarcoidosis, primary or secondary amyloid deposition, metabolic disorders like diabetes mellitus, uremia, hypothyroidism, and neoplastic disorders such as lymphoproliferative diseases, small cell lung cancer, POEMS and multiple myeloma. Routine biochemical studies were normal with the exception of low vitamin B12 level (172 pg/mL). Markers for vasculitic diseases were unremarkable. HIV and hepatitis panels were negative. In cerebrospinal fluid (CSF) examinations, there were no pathological cells, IGG index and protein levels were normal, but oligoclonal bands were pattern 2. Levels of GM1 IGM and GT1b IGM antibodies showed borderline results. Dermatological examination for pruritus revealed unremarkable results. He was first given first-line drugs in neuropathic pain. However, he could not tolerate those drugs due to various side effects. Further investigations on neoplastic, infectious and rheumatological diseases were normal. However, the patient started to have autonomic symptoms like tachycardia episodes and severe pruritus, which only responded to prednisolone 1 mg/kg/day. We administered 2 gr/kg/5 days IVIG with the diagnosis of acute demyelinating inflamatory polyneuropathy. All symptoms improved within one week and he did not have any complaints or symptoms during the 18 months follow-up.

Discussion

Acute inflamatory polyneuropathy is a monophasic disorder limiting itself within two months. The most frequent findings are stocking and glove type paresthetic sensations and weakness predominantly involving proximal parts due to radicular involvement (3,4). Although localizing symptoms and signs in our patient was a big challenge, predominant axial and proximal weakness with electrophysiological and imaging findings suggested multifocal relatively symmetric demyelinating radicular lesions. Although high CSF protein is an expected and supportive criterion for inflamatory neuropathies, normal CSF protein finding is also possible and does not exclude the diagnosis (4). All the possible etiological factors were excluded leaving the possibilities of acute demyelinating inflamatory polyneuropathy and acutely presenting chronic inflamatory demyelinating polyneuropathy. The differentiation bears importance regarding the choice of treatment. The temporal window of events at the time of admission was short; cranial nerve and autonomic system involvements were evident and antecedent infection was present. Taken together with previous reports (3), we thought that these symptoms supported the diagnosis of acute inflamatory polyradiculopathy. He did not have any additional symptoms during the 18-month follow-up period and all the previous symptoms improved completely, further supporting the diagnosis.

Neuroanatomy of itchiness consists of histamine receptor, mechanosensitive C fiber, dorsal root ganglion, medulla spinalis, thalamus, primary somatosensorial area and anterior cingulate cortex. Neuropathic itch may originate from any lesion along this pathway. Peripheral causes for neuropathic itch are cervical/lumbar radiculopathy due to tumor, disc pathologies, postherpetic neuralgia, polyneuropathy (5,6,7,8) and central causes are Creutzfeldt-Jakob disease, multiple sclerosis and neuromyelitis optica (9,10,11).

Yamaoka and colleagues reported itching in diabetic polyneuropathy patients. For those patients, they related itching to loss of sudomotor function or direct C fiber injury (12). However, this explanation does not account for the case of polyneuropathy

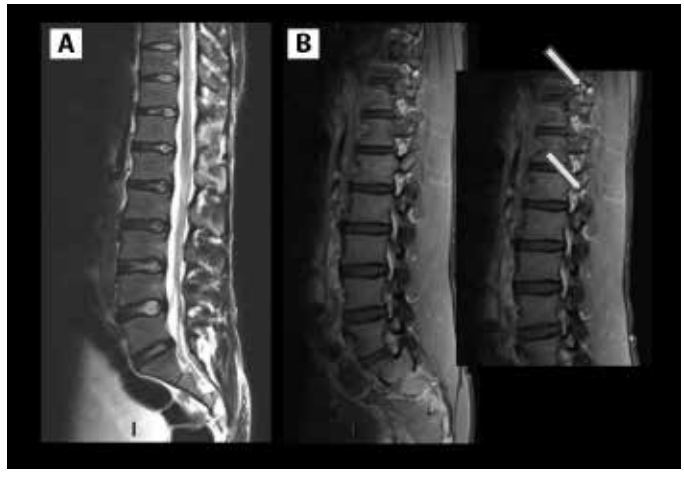

Figure 1. T2 (A) and gadolinum enhanced (B) magnetic resonance imaging demonstrating weak contrast enhancement of lumbar radices

involving large myelinated fibers like our patient. Itching closely paralleled autonomic dysfunction in our patient. There may be two explanations for the autonomic involvement. First, involvement of the myelinated part of the most proximal parts of nerves may play a role. Second, keeping in mind the discovery of antiganglioside antibodies and circulating antibodies directed against sympathetic neurons (13,14), we may speculate that a similar mechanism may be the underlying factor in the pathogenesis of itching in our patient. Another possibility is the small-fiber neuropathy. Recently, increasing numbers of Guillain-Barre patients are reported to have small-fiber neuropathy/ganglionopathy (15). However, prolonged tibial SEP and F waves decreased the possibility of pure small-fiber neuropathy in our case.

Conclusion

Acute demyelinating polyradiculoneuropathies may involve only the radices. Diagnosis of our patient was delayed as initial diplopia and pruritus complaints were considered as nonspecific and unrelated to the nervous system. We suggest that pruritus may be a remarkable neurological finding especially when it is accompanied with other neurological symptoms.

Concept: Ayşegül Gündüz, Feray Karaali-Savrun

Design: Ayşegül Gündüz, Feray Karaali-Savrun

Data Collection or Processing: Gülsün Ersen, Başak Yılmaz

Analysis or Interpretation: Ayşegül Gündüz, Feray Karaali-Savrun

Literature Search: Ayşegül Gündüz, Gülsün Ersen, Başak Yılmaz

Writing: Ayşegül Gündüz

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