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Postdural Puncture Headache and Acute Subdural Haematoma in Sjögren's Syndrome

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Abstract

Although spontaneous intracranial hypotension cases related to connective tissue diseases have been reported in the literature, to the best of our knowledge, no cases of iatrogenic intracranial hypotension have been described. In this paper, we plan to discuss a case of acute subdural haematoma and postdural puncture headache that developed after spinal anaesthesia in a patient with Sjögren's syndrome.

Keywords: Acute subdural haematoma, connective tissue disorder, postdural puncture headache, Sjögren's syndrome

Introduction

Sjögren's syndrome (SS) is a connective tissue disease, in which organ-specific or systemic symptoms may occur in addition to the exocrine gland dysfunction. The peripheral or central nervous system involvement is reported in 0%-68% of cases (1). There are case reports of cerebral infarction due to vasculopathy (2) and intracranial bleeding (3) in the literature.

One of the most common complications of spinal anaesthesia is the developing headache with cerebrospinal fluid (CSF) leakage from the extradural space following dural puncture. The needle size, needle tip, gender, pregnancy and young age are factors shown to be associated with a high risk of headache. Although there are many studies in the literature on spontaneous intracranial hypotension in connective tissue disorders, to the best of our knowledge, no study has yet been conducted on the risk of postdural puncture headache (PDPH).

This study aimed to discuss causes of PDPH and acute subdural haematoma in a case of primary SS in a scope of literature data.

Case Presentation

A 61-year-old female patient diagnosed with primary SS and bladder cancer underwent cystoscopy under spinal anaesthesia. Three hours after the operation, the patient was consulted due to throbbing headache in the back of the neck, radiating to the forehead accompanied by tinnitus, photophobia and phonophobia, with decreased severity when lying down and increased severity when standing up. The visual analogue scale (VAS) score was 7 at the initial examination. A physical examination revealed no neurological pathology aside from orthostatic headache. Biochemical results and the bleeding profile were within the normal range. According to the patient's anaesthesia record, spinal anaesthesia was performed with a 22 G atraumatic spinal needle at the L3–L4 interspace as a single application. The patient was diagnosed with PDPH. The VAS score >4 led to the administration of theophylline 200 mg (Polteofilin 200, Polifarma Pharmaceuticals) as a 45-minute slow intravenous infusion accompanied by monitorisation. After the medical treat-

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ment, the VAS score regressed to 2. A follow-up examination performed the next day showed that the orthostatic headache had passed, but the patient described a headache unrelated to the posture this time. Due to a change in the character of the pain, cranial computerised tomography (CT) for differential diagnosis was performed. The CT showed isodense, overlying bleeding in the fronto-parieto-temporal region, suggestive of subacute subdural haematoma (Figure 1). The Neurosurgery Department was consulted, and they did not suggest a surgical intervention. To prevent the bleeding risk of hypotension caused by dural leakage, an epidural blood patch was planned. After the procedure, neurological examination was repeated in intervals, and no abnormal finding developed. A follow-up CT taken 4 days later concluded that the subdural haemorrhage had regressed (Figure 2). The patient, whose headache and tomography findings had regressed, was discharged with a follow-up recommendation. There were no additional complaints at the 1-month follow-up with neurosurgery. Informed written consent was obtained from the patient for this case report.

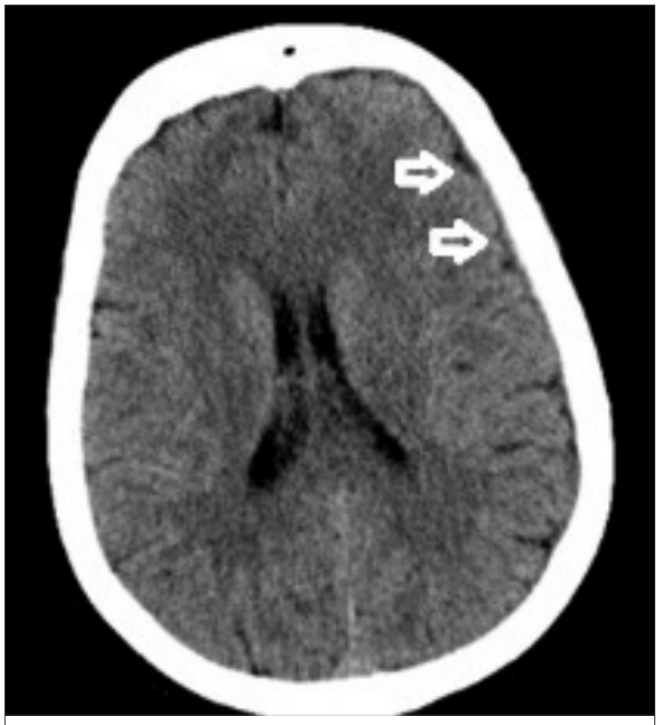

Figure 1. Subdural haematoma

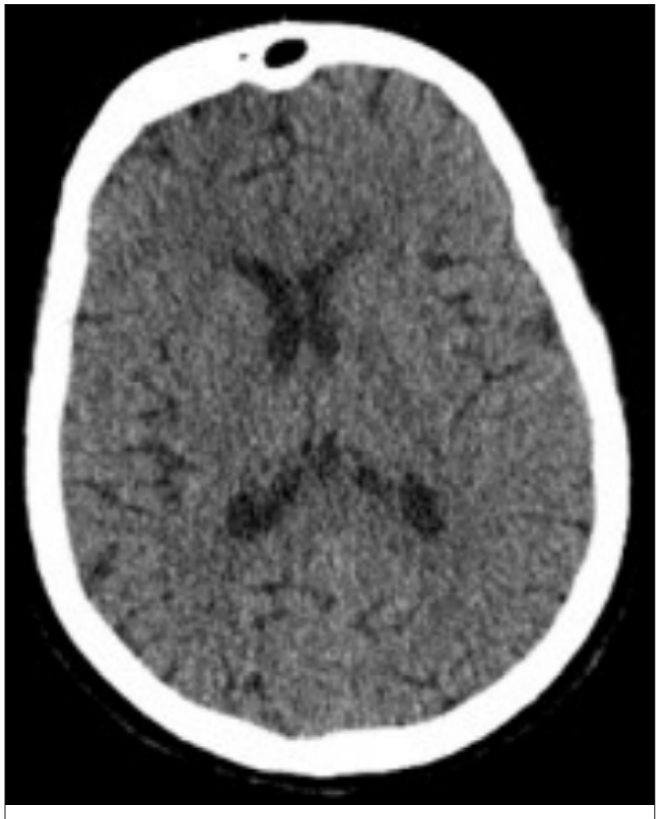

Figure 2. Follow-up computed tomography

Discussion

Intracranial haemorrhage is a serious complication for both SS (3) and PDPH (4). In our patient with SS, early PDPH followed by the diagnosis of intracranial haemorrhage raises the question of whether patients with connective tissue diseases should be considered in risky groups for these two pathologies. We believe that dural weakness in these patients may increase the amount and duration of CSF leakage and that cerebral vasculitis that may develop throughout the course of the disease may increase the haemorrhage risk of hypotension following the CSF leakage.

According to the International Classification of Headache Disorders (ICHD-3 BETA), the diagnostic criteria of PDPH include orthostatic headache, history of dural puncture and headache developing within 5 days following dural puncture (5). Our patient underwent a spinal anaesthesia procedure, reported headache that prominently changed when laying down or standing up, and was treated for PDPH. In the PDPH treatment, oral or intravenous hydration, caffeine and absolute bed rest (conservative treatment) are recommended foremost. Medical treatment including sumatriptan, adrenocorticotropic hormone, desmopressin acetate, theophylline or epidural blood patch, epidural fibrin glue or epidural saline may be applied to patients who do not benefit from conservative treatment. Surgery is the final alternative in patients who are resistant to treatment (6). In our patient, intravenous theophylline was administered in addition to conservative treatment because the VAS score was 7. A significant decrease in pain following theophylline administration was interpreted as the regression of intracranial hypotension.

There are other case reports in the literature on subdural haematoma due to PDPH (4). Yamashima et al. (7) showed that bridging veins had a thinner wall structure in the subdural

space. This suggests that displacement of the brain due to a reduced CSF volume in PDPH may cause tearing of the distended bridging veins and may lead to subdural haematoma.

The literature indicates that connective tissue diseases such as Ehler–Danlos syndrome, Marfan syndrome and isolated joint hypermobility increases the risk of spontaneous intracranial hypotension. Mokri et al. (8) identified connective tissue disease in 9 of 58 patients diagnosed with spontaneous intracranial hypotension. Another study showed that 12 (67%) of 18 patients diagnosed with spontaneous intracranial hypotension had a connective tissue disease or isolated joint hypermobility (9). A study on 50 patients diagnosed with spontaneous intracranial hypotension determined that 9 (18%) had a connective tissue disease, and 8 (16%) had isolated joint hypermobility (10). These findings may be interpreted as an indicator of the dura mater fragility in connective tissue disorders.

Hypertrophic pachymeningitis is a diffuse or local thickening of the intracranial-spinal dura mater due to inflammatory cell infiltration or interstitial fibrosis, and it may occur in intracranial hypotension (spontaneous or secondary to dural puncture), infection, autoimmune/vasculitic diseases such as SS and malignancy or it may be detected idiopathically (11). Its gadolinium contrast magnetic resonance finding is thickening of the dura mater and increased contrast retention. In Japan, between 2005 and 2009, a study that investigated 159 idiopathic hypertrophic pachymeningitis (IHP) cases found that 2 patients had SS as etiologic factors. The same study reported that the most common symptom in the 4-year period was headache (71.1%), although information on the character of the headache was not given (12). Rossi et al. (13) found dural fibrosis and the CD4 T-cell mediated inflammatory infiltration in dural biopsy of 3 patients. Nakano et al. (14) detected inflammatory infiltration of plasma cells and small lymphocytes in a dural biopsy of 3 patients diagnosed with IHP. Nevertheless, the incidence of hypertrophic pachymeningitis in SS is unknown. Closure of the dural puncture depends on the damage and fibroelastic proliferation of the dural membrane (6). We believe that inflammation of the dura and its resulting thickening may delay the recovery/closure of dural puncture. In our patient, intracranial haemorrhage was thought to be caused by CSF leakage following dural puncture, and despite the regression of symptoms with subsequent theophylline treatment, the decision to perform epidural blood patch was made.

To the best of our knowledge, there is 1 case in the literature of a patient who presented with intracranial hypotension symptoms such as positional headache, nausea, horizontal diplopia, photophobia and hypertrophic pachymeningitis and who was diagnosed with Wegener granulomatosis (15). In that case, positional headache was attributed to hygroma secondary to nontraumatic subdural haematoma. In our patient, severe orthostatic headache at the initial examination led to a delayed differential diagnosis of acute subdural haematoma. We believe that this condition should be considered in patients with connective tissue diseases.

Conclusion

We believe that the central nervous system involvement should be investigated and that an early detection of PDPH following dural puncture should lead to planned imaging for differential diagnosis of intracranial haemorrhage in patients diagnosed with connective tissue diseases such as SS.

We conclude that documentation of spinal anaesthesia procedures and ensuing complications is important in the follow-up, especially in terms of the central nervous system involvement, in patients with connective tissue diseases.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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