

CASE REPORT

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Doi:

Rhinitis as the Presenting Symptom of Pineal Region Epidermoid Tumor: A Case Report

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ABSTRACT

Brain tumors are the rarest cause of cerebrospinal fluid rhinorrhea. Non-traumatic cerebrospinal fluid rhinorrhea is also a relatively rare condition. It may be misdiagnosed as allergic rhinitis or chronic sinusitis and lead to unsuitable treatment. We described a 34-year-old man who came to our allergy clinic with a chief complaint of clear rhinorrhea from his left nostril with more than four years of duration. Only hypertrophy of left inferior concha was found in the clinical examination. His rhinorrhea aggravated when bending forward. So we were suspicious of CSF rhinorrhea. MRI was done for him and demonstrated a large tumor in the pineal region. The patient underwent surgery with resection of the mass via an infratentorial-supracerebellar approach. This case showed the role of maintaining differential diagnosis for a common complaint; rhinitis which is seen as usual.

Keywords: Brain Tumor; Cerebrospinal fluid; Rhinitis; Rhinorrhea

INTRODUCTION

Cerebrospinal fluid (CSF) leak is a condition that occurs following an improper communication or fistula between the dura and pneumatized area in the skull base like the sinusal tract or the middle ear or mastoid region.

CSF rhinorrhea can be divided base on the etiology

into traumatic and non-traumatic. Traumatic fistulae are the common causes of CSF rhinorrhea which consist of accidental (up to 80%) and post-surgical (16%).^{1,2} Non-traumatic CSF rhinorrhea is a relatively rare condition in which hypertensive obstructive hydrocephalus is formed due to an intracranial tumor.³ It may be misdiagnosed as allergic rhinitis or chronic sinusitis and lead to unsuitable treatment because of the precipitating causes which are not fully definite in most cases.² Allergic rhinitis, unlike non-allergic rhinitis, is characterized as IgE-mediated inflammation of the nasal passageways but both of them usually have

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similar symptoms, including rhinorrhea, itching, postnasal drip, congestion, and sneezing.^{4,5} Disorders can be considered as a differential diagnosis of allergic rhinitis include non-allergic rhinitis, infectious, inflammatory or immunologic, physiologic, drug-induced Rhinitis, reflex-induced, environmental factors, and NARES (non-allergic rhinitis with eosinophilia syndrome).⁴ Brain tumors very rarely presented with CSF rhinorrhea.⁶ Intracranial epidermoid cysts (ECs) are the uncommon tumor which constitutes 0.2–1% of all intracranial tumors. The most commonly located at the cerebellopontine angle, but they rarely appear in the pineal region representing 1.5–2.0% of the tumors. Intracranial EC usually presents with symptoms of parinaud's syndrome and hydrocephalus. Hemiparesis and cerebellar signs can also be noticed.^{7,8} We described the extremely rare and interesting case of non-traumatic CSF rhinorrhea with more than four years duration as a result of EC in the pineal region.

CASE PRESENTATION

A 34-year-old male came to our allergy clinic with a history of clear rhinorrhea from the left nostril for 4 years ago which increased since last year. Mashhad University has not ethics committee code for case reports. The informed consent form is obtained from the patient.

When his problem was started, he had a headache and dizziness for one week. He had no other allergic symptoms like itching, cough, sneezing. In the first-degree relatives, the history of atopy was negative. He had been referred to many physicians and managed as allergic rhinitis with antihistamine and intranasal corticosteroid but no improvement in the symptoms was observed. He had no previous history of trauma, meningitis, nasal or neurosurgical procedures. Physical examination showed hypertrophy of left inferior concha. Clear watery rhinorrhea from his left nostril was reported to become worse with positional changing such as bending forward or Valsalva's maneuver. Neurological examinations were normal. The nasal discharge was evaluated (sugar and protein) which had the chemical characteristics of CSF(table1). There was evidence of space-occupying lesion in the pineal region in brain radiologic investigation which leads to raised intracranial pressure (ICP), hydrocephalous, and CSF leakage (Figure 1). We referred the patient to the

neurosurgery department for evaluation. Endoscopic third ventriculostomy (ETV) was performed. The mass was reached at the posterior of the third ventricle but the sample did not obtain because it seems to be an epidermoid tumor. As the next step, the tumor was removed via an infratentorial-supracerebellar approach. After surgery, CSF leakage stopped, there were no neurological signs. The pathologic investigation confirmed the EC (Figure2).

Table1. CSF findings

Test name	Result
Appearance	Normal
Protein	0.35 g/L
Glucose	3 mmol/L
G serum ratio	0.6
WBC	0
RBC	0

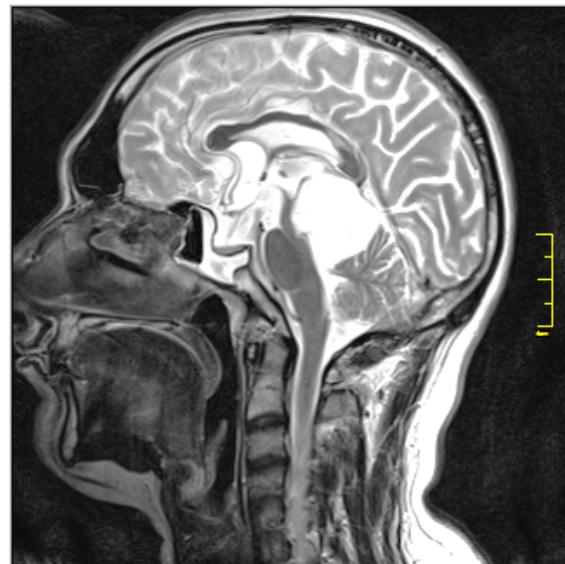


Figure 1. Preoperative Sagittal T1-weighted magnetic resonance imaging (MRI) demonstrates a large tumor in the pineal region. **Figure2:** epidermoid cyst: cystic lesion with squamous epithelium containing layered keratin material. H&E, 100X

DISCUSSION

CSF rhinorrhea is a life-threatening condition that refers to the clear watery fluid drainage from the nose that due to direct communication between the subarachnoid space and a pneumatized area of the nose and paranasal sinuses.^{1,2,9} CSF rhinorrhea may be misdiagnosed as allergic rhinitis or chronic sinusitis and lead to unsuitable treatment.

CSF rhinorrhea is a relatively rare condition compared to other rhino pusses such as allergic rhinitis, vasomotor rhinitis (VMR), and rhinosinusitis which can be classified as traumatic and non-traumatic.⁹ Trauma (80%) and surgery (16%) are the most frequent causes of CSF rhinorrhea. Non-traumatic CSF rhinorrhea is more uncommon (4%). They can be subclassified into high or normal pressure leaks, mostly they are referred to diseases that cause high ICP or local skull destruction for example hydrocephalus, tumors, osteomyelitis of the skull, and brain cysts. Furthermore, congenital defects of the skull can be the source of fistulae.^{10,11} Sometimes CSF rhinorrhea remains undiagnosed and it could lead to serious sequelae such as meningitis and possibly death.⁵ In our case, the patient's rhinorrhea was several times misdiagnosed as allergic rhinitis. ECs are benign and slow-growing tumors that comprise 0.2–1% of all intracranial tumors. Accurate pathogenesis of epidermoids, in general, is still a challenging topic. They are frequently located in the cerebellopontine cistern, followed by the parasellar cistern, but the occurrence of ECs in the pineal region are very uncommon and occurring in 10% of all pineal region tumors.⁸ Desai and colleagues reviewed 24 cases of epidermoids in the pineal region which the most common symptoms were generalized headache, ataxia, deteriorating vision, and giddiness. Parinaud's syndrome, apathy, memory disturbances, and altered behavior, drowsiness, the hemispheric symptom of hemiparesis were the other symptoms.⁸ To the best of our knowledge, this is the first case report of EC in the pineal region which presented as rhinorrhea. Biochemical tests for glucose, protein, or chloride content of the nasal discharge can help to confirm a CSF leak. CSF has a greater amount of glucose and chloride and a lower level of protein than the nasal secretions. Protein β2-transferrin examination is a gold standard confirmation of a diagnosis of CSF.¹² In our case, it was done that had the chemical characteristics

of CSF. The best methods of determination between a traumatic or neoplastic lesion and also localization of the bone defect are high resolution computed tomography (CT) and magnetic resonance (MR) imaging. Our patient was diagnosed, based on clinical examination and MRI, which showed the site of leak tumor clearly.^{10,12} Therefore, performing biochemical tests with radiologic studies at the same time is essential for diagnosis and guide management. One of the most important complications of conservative management of CSF leaks is bacterial meningitis, so surgical closure of the dehiscence is the preferred treatment to prevent ascending meningitis.¹³ The approach for repairing a CSF fistula depends on the location of leaks or defects at the skull base, the fistula size, and the flow volume. Spontaneous CSF fistulas have a lower chance to cure after surgery compared to traumatic fistulas, therefore they need more monitoring for recurrence and increased ICP. CSF leaks have been associated with a 25–87% risk of recurrence after repair.^{13,14}

Finally, the patient's position was the most important factor in this case report. No one had noticed that the patient's rhinorrhea was positional. Some factors may increase the possibility of allergies and exclude other issues such as type of secretion, pale nasal mucosa, the consistency of secretion, accompanying by polyps, response to appropriate treatment with spray cortisone and associating with other allergic diseases such as asthma and atopic dermatitis is in favor of allergens.

Stages of treatment for patients with early suspicion of rhinitis are done according to Allergic Rhinitis and Its Impact on Asthma (ARIA) Guideline and depending on the severity of the symptoms.

In conclusion, we must be concern about many conditions that mimic allergic rhinitis. Attention to the detail of history and physical examination should be considered. This case showed the role of maintaining differential diagnosis for a common complaint; rhinitis which was mundane.

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