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Settling the dispute of Romantic composer Bedřich Smetana's neurological deterioration

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Review Article

Shih-Chuan Hsiao takes responsibility for the integrity of the content of the paper

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Abstract

Background. The father of Czech music, Bedřich Smetana was a brilliant, patriotic Romantic composer who spent his last decade completely deaf. He became progressively ill in his final years and passed away prematurely at 60 years old. Since then, there have been two main propositions for the etiology of his neurological symptoms, in particular his hearing loss: neurosyphilis or osteomyelitis of the temporal bone.

Methods. This article compares the clinical presentation and pathology of neurosyphilis and osteomyelitis.

Results. This article infers which one is arguably the most likely cause based on Smetana's own medical history, signs and symptoms and autopsy findings.

Conclusion. Smetana's clinical presentation and pathological results grant us a clearer picture of his neurological condition and allows us to diagnose his final neurological deterioration as complications of neurosyphilis and not osteomyelitis of the temporal bone.

Background

Bedřich Smetana (1824–1884) of the Romantic Era is known today as the father of Czech music.[1] Famous works of this patriotic composer include Má vlast (My Fatherland), Prodaná nevěsta (The Bartered Bride), and String Quartet No. 1 in e minor: Z mého života (From My Life).[2] Gifted from a young age, Smetana flourished as a composer despite spending the last 10 years of his life deaf.[3] His life's work was tragically cut short as he then became progressively ill until his untimely death at 60 years of age in a mental asylum in Prague.

Clinical presentation

Smetana's medical history and notable musical works throughout his life are summarized in Table 1.[1,2,4–9]

Smetana was born on 2 March 1824. In his medical history, he was afflicted with osteomyelitis of the right temporal bone and mandible due to a gunpowder explosion in his childhood.[1,2,4]

Smetana's illness first manifested itself in the form of tonsillitis in 1874 and subsequent general exanthema, dizziness, and the catastrophic symptoms of tinnitus, auditory hallucinations, and progressive bilateral hearing impairment until complete deafness within a few months.[1,2,4–6] He was afterwards diagnosed with "labyrinth paralysis" in 1875 and treated with politzerisation, electrical stimulation, and mercury ointment, all of which failed. In 1879, he also developed symptoms of depression and what he himself suspected was insanity.

From 1881 onward, Smetana's condition deteriorated sharply both physically and mentally, and his symptoms persisted until his death in 1884.[2] He was hypersensitive to coldness and had an ongoing cough with severe dyspnea, nocturnal chest pain, and crackles.[5] In addition to confusion, memory deficits and mental exhaustion, he also had repeated visual and auditory hallucinations, along with ataxia and expressive aphasia, which were exacerbated after a stroke-like seizure in 1882.[1,6] In his final year, the composer had fully developed dementia, unable to recognize family or friends, and was psychotic with unpredictable agitation and rage attacks. He also had left facial paresis and swallowing difficulties. He was severely cachectic, and his speech and letters were incomprehensible. Smetana passed away on 12 May 1884.

Table 1. Medical history and notable musical achievements of Bedřich Smetana, born 2 March 1824 and died 12 May 1884

Year	Medical history		Notable compositions
	Birth of Bedřich Smetana: 2 March 1824		
1835	• Osteomyelitis of right temporal bone & mandible due to an explosion of a glass bottle of gunpowder		
1848–1849			• Festive Overture in D Major
1853–1854	• 1854: Death of daughter Gabriela (1852–1854) due to tuberculosis		• Festive Symphony in E Major
1855–1857	• 1855: Death of daughter Bedřiška (1851–1855) due to scarlet fever • 1856: Death of daughter Kateřina (1855–1856) due to tuberculosis • 1857: Start of intimate relationship with student Fröjda Benecke		• Piano Trio in g minor (1855)
1858–1861	• 1859: Death of wife Kateřina Kolářová (1827–1859) due to tuberculosis		• Richard III • Wallenstein's Camp • Hakon Jarl
1863			• The Brandenburgers in Bohemia
1866			• The Bartered Bride
1868			• Solemn Prelude in C Major • Dalibor
1874	• Suppurative ulcer, with profuse exanthema 3 months later • Severe angina (tonsillitis / angina tonsillaris) → • Attacks of dizziness, progressive & persistent tinnitus, progressive bilateral hearing loss until complete deafness, auditory hallucinations → Tinnitus increased when composing • Attempted (failed) treatment of ear with politzerisation (Eustachian tube inflation)		• Má vlast (1874–1879, completed after loss of hearing) • The Two Widows
1875	• Diagnosed with 'labyrinth paralysis' (with central impairment & neuritis) • Unsuccessful treatment with electrical stimulation & mercury treatment (with mercury ointment Schmierkur)		
1876			• String Quartet No. 1 in e minor: Z mého života • The Kiss
1877	• Retain strong ability to compose music despite illness (music composition used as escape from troubles in life)		
1878	• Wife Barbora Ferdinandiová & children infected with measles		• Czech Dances • The Secret
1879	• Symptoms of depression, with: melancholic & constricted feelings, mental exhaustion, constant brooding over 'misfortune' of hearing loss, difficulty concentrating for longer periods of time (>1 h) • Suspected insanity by Smetana himself		
1881		(persistent & progressive until death in 1884)	• Libuše
1882	• Cough with severe trouble breathing, chest pain at night, & rales • Flickering vision • Stroke-like seizure • Cold intolerance	• Changed mental status, with confusion, memory deficits, & mental exhaustion • Repeated visual & auditory hallucinations	• The Devil's Wall • String Quartet No. 2 in d minor (1882–1883)
1883		• Speaking difficulties & repeated episodes of expressive aphasia, especially after stroke-like seizure in 1882	• Prague Carnival • Viola (incomplete)
1884	• Dementia, with inability to recognize family or friends • Psychotic, with unpredictable agitation & rage attacks • Left facial paresis, with difficulty swallowing • Severe cachexia	• Incomprehensible speech & incoherent letters • Gait abnormalities, incl. ataxia	
	Death of Bedřich Smetana: 12 May 1884		

Autopsy

Two autopsies have been performed on Smetana's body, in 1884 immediately post-mortem and a century later in 1987 when the body was exhumed and re-examined.[1,2,4,7] The findings of both revealed:

- skull asymmetry, with hypoplasia of middle and lower third of right facial skeleton
- no significant inflammatory changes in right temporal bone or mastoid process
- (microscopic) signs of inflammation in right malleus and incus
- bilateral cochlear nerve atrophy without major pathological changes of inner ear
- chronic internal hydrocephalus, with ventricular dilation
- cerebral atrophy, with iron deposition in brain cortex
- granular ependymitis
- chronic leptomeningitis, with meningeal thickening and *Treponema pallidum* found in brain
- positive serology for syphilis in various regions of the body (*Treponema pallidum* haemagglutination test (TPHA) +,

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fluorescent treponemal antibody-absorption test (FTA-ABS IgG) ±, rapid plasma regain (RPR) test +++)
- chronic endarteritis, with suspected atherosclerotic or cerebral vascular sclerotic change
- thrombosis arteriae femoralis
- bilateral lobar pneumonia
- furunculosis
- block vertebrae at C2/3
- huge concentrations of mercury in tissues

Discussion

As medicine and technology were not as advanced or well-developed in Smetana's time as they are today, doctors could only rely on history taking, physical examination, and post-mortem examinations to make a diagnosis. Smetana's doctors therefore concluded with logical reasoning that he had labyrinth paralysis and treated his condition symptomatically.[1,4] From a modern point of view, however, and based on his autopsy results, Smetana died of neurosyphilis complicated with bilateral lobar pneumonia.

Today, hearing loss can be classified into two major groups: conductive hearing loss, in the outer and/or middle ear, and sensorineural hearing loss, in the inner ear, cochlear nerves, and/or brain. It can also be divided into lesions of the central or peripheral nervous system. Since Smetana's death, two main potential etiologies for his deafness and other debilitating neurological symptoms have emerged based on his clinical presentation and the autopsies.[1,4,7] One is that the composer suffered from complications of osteomyelitis of the right temporal bone. The other is that he was infected with syphilis, which progressed to neurosyphilis.

Osteomyelitis of the temporal bone is a bone infection that can be caused externally through trauma or medical procedures or internally as a complication of mastoiditis.[10] Mastoiditis is often a complication of acute or chronic otitis media with an internally disseminated infection that destructs the mastoid bone. Possible symptoms include local inflammation or tenderness, (chronic) ear discharge, and general complaints like headache, exhaustion, or fever. Both osteomyelitis and mastoiditis can result in further sequelae such as (conductive) hearing impairment or cranial nerve dysfunction. They are treated symptomatically and, when needed, with antibiotics or surgical intervention.

Neurosyphilis is an infection of the central nervous system (brain and/or spinal cord) by *Treponema pallidum* that can occur especially if a previous syphilis infection has not been thoroughly treated.[11,12] Complications include visual and/or auditory hallucinations, as well as meningeal or meningovascular syphilis, which may lead to meningitis or ischemic stroke. General paresis usually affects the frontotemporal brain parenchyma and causes changes in the patient's mental status. Tabes dorsalis, demyelination of the dorsal columns and dorsal root ganglia, results in impaired proprioception and therefore ataxia. In later stages, damage to lower cranial nerves could also result in manifestations like facial paralysis and swallowing difficulties. If the infection spreads to the ear, it can also induce (sensorineural) hearing loss, tinnitus and dizziness. The mainstay of treatment for syphilis is penicillin.

In Smetana's case, his clinical presentation and the two autopsies clearly indicate that the problem lay in the central nervous system.[1,2,4,7] Complete deafness, tinnitus, hallucinations, change in mental state and gait abnormalities all coincide with neurosyphilis. The positive serology for syphilis, along with proof of meningeal inflammation and cerebral changes in the autopsy, also validate the diagnosis. This is supported by records of his personal life, as he had at least one known mistress and lived an active social life.[3]

Furthermore, Smetana's accounts and the pathological results argue against osteomyelitis or a peripheral lesion as the cause for his deafness. No significant pathological changes have been found in the middle or inner ear on either side, and the ossicles did not suffer severe damage despite his asymmetric skull.[4] Smetana did not complain of other signs of chronic otitis media such as long-term ear discharge, infection or inflammation,[1] so it was unlikely that he developed chronic otitis media from tonsillitis. It was also unlikely the rapidly progressive bilateral hearing impairment that first appeared in 1874 was caused by the right-sided osteomyelitis that happened 40 years prior.

There was likewise a low probability that Smetana suffered from adult meningitis as a possible complication of childhood temporal bone fracture.[13,14] Posttraumatic meningitis was not a prime consideration, as Smetana did not complain of chronic otorrhea or rhinorrhea or other potential signs or symptoms of a posttraumatic CSF leak.[15] The onset of his neurological symptoms first occurred about 40 years after his facial injury from the gunpowder explosion (Table 1), and no fractures were documented in either Smetana's records or the autopsies.[1,2,4,7,16] Rather, the osteomyelitis and subsequent hypoplasia of the right facial skeleton were a result of bone tissue damage caused by a complicated soft tissue infection from the injury.[16]

In addition to neurosyphilis, Smetana might have suffered from mercury poisoning, based on the high concentrations of mercury discovered in his body.[1,17] Although syphilis had never been confirmed while he was alive, mercury was a common first-line treatment for syphilis in the 19th century. It is also known today that mercury is a neurotoxin that could lead to psychosis and slow down the auditory brainstem response, which may have aggravated Smetana's neurological symptoms.

- Smetana was a renowned Romantic composer whose loss of hearing in his final decade has remained ambiguously explained for the past century.
- Smetana's hearing loss was most likely caused by neurosyphilis.
- Smetana's clinical picture overwhelmingly favors syphilis over osteomyelitis.
- Mercury poisoning may have accelerated Smetana's hearing loss.
- As supported by his clinical presentation and autopsy results, the authors conclude that Smetana died of neurosyphilis complicated with bilateral lobar pneumonia.

Conclusion(s)

The true nature of Bedřich Smetana's illness has been under debate since his death over a century ago. From Smetana's clinical presentation to pathological analyses from autopsies, we have a clearer picture regarding the etiology of his neurological condition in the last decade of his life. Were Smetana alive today, he would have most likely been promptly diagnosed with syphilis and treated with modern medical resources.

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