

Comment

From ear to brain

The pachymeninx is referred to as the dura mater, while the leptomeninges are the thinner pia mater and arachnoid mater membranes. As a rule, all three meninges are attacked in infectious meningitis, while the dura mater is primarily affected by inflammatory meningitis (1). Sarcoidosis, Sjögren's syndrome, Wegener's granulomatosis, temporal arteritis, systemic lupus erythematosus and rheumatoid arthritis are examples of diseases that can cause inflammatory meningitis besides the idiopathic form of pachymeningitis. Pachymeningitis is regarded as being associated with immune-induced changes in the dura mater. All three meninges become connected together by fibrous adhesions in the further inflammatory process.

The patient in question had a complicated history of illness. He was assumed to have intracranial hypertrophic pachymeningitis, probably caused by Wegener's granulomatosis. In my view, there might have been another cause.

The patient had had recurrent otitis with secretion from the left ear. Repeated MRI of the cerebrum and CT of the temporal bones revealed opacity of the pneumatization in the left temporal bone. There was fluid and soft tissue congestion around the internal carotid artery and internal jugular vein below the base of the skull. During down-titration of corticosteroid therapy, the patient developed right-side external ophthalmoplegia and numbness corresponding to the ophthalmic branch of the trigeminal nerve. This can be related to the changes in and around the apex of the petrous portion of the temporal bone on the opposite side.

In 1904, Gradenigo described a series of patients with acute or chronic middle ear

inflammation, paresis or paralysis of the abducens nerve and trigeminal neuralgia, corresponding in particular to the ophthalmic branch (2). The term Gradenigo's syndrome was later associated with the triad of symptoms. Gradenigo believed that the neurological symptoms were a result of local leptomeningitis triggered by the ear infection. The same year, Citelli maintained that the disease picture was due to pachymeningitis that was a result of infection in the petrous apex of the temporal bone. In 1905 a third Italian, Dorello, described a non-elastic canal at the apex of the petrous temporal bone (which is still named after him), through which the abducens nerve runs, and in close relation to the trigeminal nerve ganglion (2). In 2007, Bravo et al. described a case where middle ear inflammation with opacity of the air cells in the temporal bone could be the cause of diffuse pachymeningitis (3). The ear infection was a type of secretory otitis with vague symptoms, an extended course and without detection of an infectious agent (3).

The effectiveness of immune-modulating therapy provides support for the theory of immune mediation of pachymeningitis. The effectiveness of methotrexate has been seen in other similar case histories (3–5). This patient was planned to have maintenance therapy with methotrexate after treatment with intravenous cyclophosphamide. After completion of treatment with cyclophosphamide, the meninges were found to be increasing in thickness, and rituximab was administered instead. The use of monoclonal antibody (rituximab) outside the approved indication requires great caution and close monitoring. Such use may be a last

resort and contributes to innovation in clinical practice, but at the same time it entails a special responsibility.

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References

1. Jones SE, Belsley NA, McLoud TC et al. Rheumatoid meningitis: radiologic and pathologic correlation. *AJR Am J Roentgenol* 2006; 186: 1181–3.
2. Felisati D, Sperati G. Gradenigo's syndrome and Dorello's canal. *Acta Otorhinolaryngol Ital* 2009; 29: 169–72.
3. Bravo D, Machová H, Hahn A et al. Mastoiditis complicated with Gradenigo syndrome and a hypertrophic pachymeningitis with consequent communicating hydrocephalus. *Acta Otolaryngol* 2007; 127: 93–7.
4. Bosman T, Simonin C, Launay D et al. Idiopathic hypertrophic cranial pachymeningitis treated by oral methotrexate: a case report and review of literature. *Rheumatol Int* 2008; 28: 713–8.
5. Voller B, Vass K, Wanschitz J et al. Hypertrophic chronic pachymeningitis as a localized immune process in the craniocervical region. *Neurology* 2001; 56: 107–9.

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