# Idiopathic hypertrophic cranial pachymeningitis (IHCP)

see also Idiopathic chronic hypertrophic craniocervical pachymeningitis.

Idiopathic hypertrophic pachymeningitis in the cranial region is an uncommon disease of unknown etiology characterized by thickening of the cerebral dura mater with possible associated inflammation.

Hypertrophic pachymeningitis is an uncommon disorder that causes a localized or diffuse thickening of the dura mater.

It was first described by Charcot and later by Naffziger and Stern [1].

## Clinical features

The most frequently described clinical symptoms include headache, cranial nerve palsy, and cerebellar dysfunction. Epilepsy and/or status epilepticus as main presentation is very uncommon.

## Case reports

### 2016

Hahn et al., retrospectively examined twenty-two consecutive cases of HP seen in a single practitioner neurology practice over a ten-year time period. The most common etiologies were idiopathic HP and neurosarcoidosis. No imaging features were completely specific to any etiology. Nonetheless, idiopathic HP typically demonstrated diffuse regular enhancement whereas neurosarcoidosis was more likely to display a nodular enhancement pattern. Headache and cranial neuropathies were the most common clinical presentation. HP symptoms were often responsive to steroids but complete responses were rare. HP is a diagnostic challenge without specific findings on minimally or non-invasive diagnostic studies. Biopsy is often required and serves as the basis for effective therapy [2].

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Two consecutive cases are presented of patients manifesting focal status epilepticus secondary to IHCP, with clinical, laboratory [blood test and cerebrospinal fluid (CSF) analysis], neuroradiologic [magnetic resonance imaging (MRI) at 3 Tesla and digital subtraction angiography (DSA)], and therapeutic data. One patient underwent meningeal biopsy; pathology findings are also included. Corticosteroid therapy resulted in clinical improvement in both cases, and neuroimaging showed decreased abnormal morphology, compared to initial findings.

In the diagnostic approach to focal status epilepticus or epilepsy, IHCP must be considered a potential, although extremely infrequent, cause. Anti-inflammatory treatment is an effective addition to antiepileptic drug therapy in patients with IHCP [3].

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A 26-year-old male presented with a 6-day history of paroxysmal headache which was worsen with

nausea and vomiting for 1 day. Head CT on admission revealed left chronic subdural hematoma with midline shift. An emergency Burr hole drainage for hematoma was performed. Headache recurred 6 days later. MRI of the brain revealed a diffuse thickening and a gadolinium-enhancement of the falx, cranial dura mater and tentorium cerebelli on the left side with pia mater involved. Lumber puncture showed increased intracranial pressure and elevated IgG level in cerebrospinal fluid. Histological examination of the biopsy specimen showed thickened, fibrotic dura with a sterile chronic inflammation. According to pathological examination, idiopathic hypertrophic cranial pachymeningitis was considered as the final diagnosis. Symptoms were improved with steroid pulse therapy [4].

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Shetty and Kadambi from Narayana Hrudayalaya, Bangalore published a case of idiopathic hypertrophic cranial pachymeningitis [5].

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A case of headache and idiopathic hypertrophic cranial pachymeningitis drastically improved after CSF tapping [6].

**2015**

A previously healthy 63-year-old man presented with a 2-weeks history of diplopia without headache. Neurological examination revealed total external ophthalmoplegia of the left eye and limitation of abduction of the right eye. Initial cranial MRI showed thickening and enhancement of the dura mater only on the anterior cranial fossa but unremarkable on the cavernous sinus. Idiopathic hypertrophic cranial pachymeningitis was diagnosed in the absence of demonstrable underlying infective, neoplastic, or systemic autoimmune disease by his clinical findings, laboratory tests and radiological examinations. Corticosteroid therapy was initiated with methylprednisolone (1,000 mg/day for 3 days), followed by oral prednisolone and tapering off. Eye movements improved with treatment and completely recovered within 4 weeks after starting administration, and cranial MRI at the 15 days after starting treatment showed improvement. Teramoto et al., suggest that his ophthalmoplegia was caused by the inflammation of dura on the cavernous sinus beyond the thickening lesion of cranial MRI. In a case of bilateral ophthalmoplegia with or without headache, it is required to examine the dural thickening and enhancement on the anterior cranial fossa by cranial MRI [7].

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Auboire et al., present the first case report of hypertrophic pachymeningitis revealed by isolated occipital neuralgia.

Idiopathic hypertrophic pachymeningitis is a plausible cause of occipital neuralgia and may present without cranial-nerve palsy. When occipital neuralgia is not clinically isolated or when a first-line treatment fails, another disease diagnosis should be considered. However, the cost effectiveness of extended investigations needs to be considered [8].

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A case of idiopathic hypertrophic pachymeningitis (IHP) forming a central skull base mass to illustrate

the process required when one investigates such skull base lesions. This is the first description of mass forming or tumefactive IHP extending into the nasopharynx. A 32-year old woman presented with frontal headaches and nasal discharge. She then deteriorated and was admitted with worsening headaches, serosanguinous nasal discharge and bilateral ophthalmoplegia. Multimodality imaging confirmed a destructive central skull base soft tissue mass involving the posterior clivus, floor of sphenoid sinus, nasopharynx and extending into both cavernous sinuses. Unfortunately, the patient continued to deteriorate despite treatment with broad-spectrum antibiotics. Cerebrospinal fluid, blood tests and transnasal biopsies for histology and microbiology did not reveal a diagnosis. Further neuroimaging revealed extension of the mass. Early corticosteroid treatment demonstrated radical improvement although an initial reducing regime resulted in significant rebound deterioration. She was stable on discharge with slowly reducing low dose oral prednisolone and azathioprine. We discuss the complexity of this case paying special attention to the process followed in order to arrive at a diagnosis of idiopathic hypertrophic pachymeningitis based on both the clinical progression and the detailed analysis of serial skull base imaging. Knowledge of the potential underlying aetiologies, characteristic radiological features, common pathogens and the impact on blood serology can narrow the potential differentials and may avoid the morbidity associated with extensive resective procedures [9].

**2014**

Takeuchi et al., report the tenth case of IgG4-related intracranial hypertrophic pachymeningitis and review the previous literature. A 45-year-old male presented with left-sided focal seizures with generalization. Magnetic resonance imaging (MRI) revealed a diffuse thickening and enhancement of the right convexity dura matter and falx with focal nodularity. The surgically resected specimens exhibited the proliferation of fibroblast-like spindle cells and an infiltration of mononuclear cells, including predominantly plasma cells. The ratio of IgG4-positive plasma cells to the overall IgG-positive cells was 45% in the area containing the highest infiltration of plasma cells. On the basis of the above findings, IgG4-related sclerosing disease arising from the dura mater was suspected. IgG4-related sclerosing disease should be added to the pachymeningitis spectrum [10].

**2005**

A 36-year-old woman was admitted to our hospital in August 2001 with a complaint of motor weakness in her left leg for 1 month. Magnetic resonance imaging with gadolinium revealed enhancement of the thickened dura mater and perifocal edema in the right frontoparietal region suggesting a mass lesion. Histological examination of the biopsy specimen revealed a dense fibrous cellular tissue. There was no obvious inflammatory infiltrate but in some areas between fibrous bundles one or two lymphocytes were detected. This is the second report in the literature of IHCP with focal edema causing a mass effect [11].

**2002**

Three cases of this entity are described. Presenting features were headaches and cranial neuropathies in two patients and predominantly cerebellar dysfunction in the third. One of the patients also had evidence of spinal involvement. Lower cranial nerves were chiefly involved in two patients whereas optic nerve was the predominantly affected nerve in one. Except for the presence of rheumatoid arthritis in one of the patients, we could not document clinical or biochemical evidence of

any predisposing infective, inflammatory or infiltrative condition in the other two. All three patients had characteristic changes on imaging suggestive of thickened and enhancing duramater. Although variable steroid responsiveness was seen in all the three patients, tendency towards steroid dependence was evident. The clinical presentations, causes, radiological features, management options and differential diagnosis of this unique clinical syndrome have been discussed [12].

### 1999

In six patients, the main manifestations were cranial nerve palsies and headache. Three associations were present, namely optic neuropathy, Tolosa-Hunt syndrome, and diabetes insipidus. Gadolinium-enhanced magnetic resonance imaging was diagnostic, showing intense dural enhancement in a linear or nodular pattern. The responses to corticosteroid therapy were better for patients who exhibited linear, rather than nodular, dural enhancement. For one patient, surgical decompression of the superior orbital fissure provided lasting relief. The course of the disease followed one of three patterns, i.e., sustained remission, relapse with corticosteroid independence, or relapse with corticosteroid dependence. Pulse corticosteroid therapy provided significant relief, while reducing the daily corticosteroid requirement and avoiding side effects, for a corticosteroid-dependent relapsing patient.

Idiopathic hypertrophic cranial pachymeningitis exhibits varied clinical courses. It is important to prevent irreversible cranial neuropathy during the active phase of the disease, using daily administration of corticosteroids, pulse corticosteroid therapy, or surgical decompression [13].

### 1997

Four patients with idiopathic hypertrophic pachymeningitis who had varied clinical presentation. Imaging studies revealed diffuse thickening of the pachymeninges; in one patient there was extensive dural sinus thrombosis. Since no identifiable cause was found, the cases were labelled as idiopathic [14].

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Three cases of idiopathic hypertrophic cranial pachymeningitis are presented. The diagnosis was based on the CT scan or MRI findings (or both) of thickened enhancing dura. In all cases, meningeal biopsies were performed and microscopic findings were compatible with nonspecific inflammation. All cases presented with subacute and chronic localized headache. Two cases had associated chronic meningitis. One cases presented with a syndrome of multiple cranial nerve involvement (polyneuritis cranialis). Corticosteroids, in the form of prednisolone 60 mg/day, were effective in all cases. Two cases with less severe pachymeningitis received corticosteroids for 2 weeks, then were tapered off in 4 to 6 weeks. A case with extensive lesions needed a long-term low dosage of prednisolone, 5 to 10 mg/day for maintenance therapy. Idiopathic hypertrophic cranial pachymeningitis may be related to the Tolosa-Hunt syndrome, the syndrome of polyneuritis cranialis, and multifocal fibrosclerosis [15].

### 1996

A successful direct operation in a case of hypertrophic cranial pachymeningitis (HCP) is described. A 51-year-old male was admitted with right visual disturbance. CT scanning revealed a mass lesion in the right middle cranial fossa extending into the right orbita. From MRI it was ascertained that this mass lesion consisted of thickening of the skull base dura. The effect of steroids was transient and his visual disturbance was rapidly increasing. A direct operation was performed consisting of partial dissection of the thickening dura and decompression of the right optic nerve. Histological diagnosis showed HCP. After surgery his symptoms dramatically improved. The etiology, natural course and treatment of HCP are well reported in the literature. in the present case the etiology was not clear, but there was a possible relationship with intraorbital pseudotumor or multifocal fibrosclerosis. The reported occasional transitory effect of steroids on HCP and a poor prognosis for visual acuity are common traits in these patients. The authors thus feel that a direct operation should be immediately carried out when the effect of steroids on visual disturbances associated with HCP is judged transient [16].

### 1993

Seven patients with cranial pachymeningitis of unknown origin in whom the main clinical features were headaches, ataxia, and cranial nerve palsies. CSF showed inflammatory changes. CT and MRI showed thickening of the falx and of the tentorium. The clinical course was chronic. Four patients improved with prednisolone but became steroid-dependent: in two cases, radiotherapy had no lasting improvement and in one, azathioprine permitted a reduction of the corticosteroids. Five patients had biopsy of the tentorium cerebelli or of the temporal dura mater. In two cases, autopsy revealed extensive pachymeningitis without parenchymal changes. In all instances, microscopic examination of the dura mater showed a cellular infiltrate of polymorphic cells; there were no epithelioid granulomas. Review of the literature discloses seven similar cases. We discuss the relationship of these lesions with inflammatory meningeal masses, the focal pachymeningitis of the Tolosa-Hunt syndrome, and multifocal fibrosis [17].

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Three cases of hypertrophic cranial pachymeningitis. These lesions typically cause progressive cranial nerve palsies, headaches, and cerebellar dysfunction. They occur in patients of all age groups; the peak incidence is in the sixth decade. Hypertrophic cranial pachymeningitis is best identified by magnetic resonance imaging. The diagnosis is established by excluding all other granulomatous and infectious diseases. A dural biopsy is essential to confirm the diagnosis. Hypertrophic cranial pachymeningitis is initially responsive to steroid therapy, but in most cases it recurs or progresses despite treatment. Surgical excision of granulomas is occasionally necessary to alleviate a mass effect. The long-term outcome remains uncertain for most patients, but progressive disease is usually fatal owing to cranial neuropathies [18].

### 1989

Three patients with diffuse idiopathic cranial pachymeningitis with predominant involvement of the tentorium and falx are reported. Progressively increasing headaches were the usual symptoms, along with ataxia and various cranial nerve palsies. CT in all cases and MR imaging in two cases detected isolated thickened dura mater. In one case, MR depicted dural involvement as a very large, hypointense area with fine hyperintense edges on T2-weighted images. Microscopic examination of thickened dura revealed extensive fibrotic tissue with a chronic inflammatory infiltrate containing

lymphocytes, plasma cells, and scattered eosinophils; these findings closely paralleled the MR features. Only four cases with similar pathologies have been described, all before the advent of CT and MR. We discuss the different causes of thickened dura mater as well as the significance of the fact that dural thickening can be responsible for occlusion of the dural sinuses. Cranial pachymeningitis is a rarely reported disease that can resemble other disorders associated with tentorial thickening; CT and MR can help differentiate it from these other disorders [19].

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