

ARACHNOID CYST



"The Card Players" oil on canvas, 1851, Adolf Tidemand

...They saw heads neatly fixed on poles being brought to the Sultan's palace. Such sights caused an increase in their argumentative discussions, and when they did not argue, boredom hung so heavily on them that the old lady bluntly asked one day:

"I should like to know which is worse, to be raped a hundred times by negro pirates, to have one buttock cut off, to run a Bulgar gauntlet, to be flogged and hanged in an autoda-fe, to be dissected, to row in a galley, to experience - finally - all the miseries we all have endured, or simply to stay here with nothing to do?"

"That's quite a question", said Candide.

Her asking the question created a host of new viewpoints, and Martin in particular, decided that men were born to live either amid the convulsions of uncertainty or in the lethargies of boredom. Candide did not agree, but he was not totally certain. Pangloss admitted to having been afflicted with constant suffering - but once having declared that everything was wonderful, he'd go on declaring it, though he didn't anymore believe a word he was saying.

There was one thing that served to confirm Martin's detestable principles, to make Candide hesitate more than ever, and to embarrass Pangloss. What happened was, one day, they saw Paqeutte and Father Giroflee come to the farm, obviously extraordinarily destitute. It had not taken them long to consume their three thousand in gold; they had separated, come back together, they had quarreled, gone to prison; they had escaped; and in the end, Father Giroflee had indeed turned Turk. Everywhere they went, Parquette practiced her profession, and no longer earned anything from it.

"I warned you", Martin told Candide, "that your gifts would soon be dissipated and would only make them more miserable. You've thrown away millions, you and Cacambo, and you're not any happier then Parquette and Giroflee".

"Ah, ha!" said Pangloss to Parquette. "Heaven brought you here among us, my poor child! Do you realize I lost the tip of my nose because of you, and an ear and one eye? What have you come to! And what a world this is!!"

This new episode led them to philosophize more than ever.

There was a famous dervish in the neighborhood who was considered the best philosopher in Turkey. They went to consult him; Pangloss was the spokesman and said:

"Master, we've come to ask you why such a strange animal as man was ever created".

"Mind your own business", said the dervish. "Is this any of your concern?"

"But, Reverend Father", said Candide, "the world is full of terrible evils".

"What difference does it make", said the dervish, "whether there's evil or good? When His Majesty sends a ship to Egypt, does he worry whether the mice in that ship are comfortable"?

"So what should we do?", said Pangloss

"Shut your mouths," said the dervish.

"I had fancied", said Pangloss, "we might have some small discussion with you about causes and effects, and the best of all possible worlds, the origins of evil, the true nature of the soul, and pre-existent harmony".

At these words, the dervish slammed the door in their faces.

While they were conducting this conversation, it became known that, in Constantinople, two ministers of state and a teacher of Muslim law had been strangled, and a number of their friends had been impaled on sharp stakes. For some hours, this dreadful event was discussed everywhere. Pangloss, Candide, and Martin, as they were returning to the farm, met an old man taking a breath of fresh air in front of his house, under an arbor of orange trees. Pangloss, who was as inquisitive as he was argumentative, asked him the name of the Islamic teacher just strangled.

"I don't know anything about that", said the good old man, "nor have I ever known the name of any Muslim teacher or minister of state. I know absolutely nothing about this business. My assumption has always been that those who meddle in public affairs often meet a miserable death, and deserve it. I never enquire into anything that happens in Constantinople. It's enough to me to have the produce of my fields sent there and sold there".

Having spoken these words, he invited the three strangers into his house. His two daughters and two sons gave them several varieties of sherbet, made with their own hands, and beverages flavoured with lemon-peel preserves, oranges, lemons, limes, pineapples, pistachios, and Mocha coffee - but not blended with wretched coffee from Batavia and the Indonesian islands. And then this good Muslim's daughters perfumed the beards of Candide, Pangloss, and Martin.

"You must be the proprietor", said Candide to the Turk, "of a vast and magnificent estate?"

"I own only twenty acres", the Turk replied. "I cultivate them with my children. Our labor keeps us from those three great evils, boredom, sin and want".

Going home to his farm, Candide thought deeply about the Turk's words. He said to Pangloss and Martin:

"I think this good old man has a better life than any of the six kings with whom we've had the honor of breaking bread".

"Greatness". Said Pangloss, "is very dangerous, according to all the philosophers, since in the end; Eglon, Moabite king, was assassinated by Ehud; Absalom was hung by his own hair and run through by three spears' King Nadab, Jeroboam's son, was killed by Baasha, King Elah by Zimri, Ahaziah by Jehu, Athaliah by Jehoiada. Kings Joachim, Jeconiah, and Zedekiah became slaves. You are familiar, are you not, with how Croesus perished, and Astyages, Darius, Denys of Syracuse, Pyrrhus, Perseus, Hannibal, Jugurtha, Ariovistus, Caesar, Pompey, Nero, Otho, Vitellius, Domitian, Richard the Second of England, Edward the Second, Henry the Sixth, Richard the Third, Mary Stuart, Charles the First, the three French Henrys, Emperor Henry the Fourth? You know...."

"I also know", said Candide, "that we need to work our fields".

"You're right" said Pangloss, "because when men were placed in the garden of Eden, they were set there ut operaretur eum - that is, so they might work. This proves that men are not born for resting".

"Let us work without thinking", said Martin. "That is the only way to make life bearable".

All the members of their small society participated in this praiseworthy plan, each working at what he or she could do best. Their small farm produced a great deal. Cunegonde was truly very ugly, but she made excellent pastry. Paquette did embroidery; the old woman was in charge of their linens. Even Father Giroflee made himself useful: he was an excellent carpenter, and actually became a respectable man. And Pangloss would sometimes say to Candide:

"Everything that happens, in this best of all possible worlds, is linked to everything else, because when it comes down to it, if you hadn't been driven out of a mansion, by savage kicks in the backside, on account of your love for Miss Cunegonde and you hadn't been taken by the Inquisition, and gone all over South America on foot, and hadn't run the Baron through with a sword, and hadn't lost all your wonderful Eldorado sheep, you wouldn't be eating here and now, lemon-peel preserves and pistachios"

"That's well said", replied Candide, "but we need to work our fields".

Voltaire, "Candide Or Optimism", 1759.

Well over two and a half centuries after it was written, Voltaire's "Candide" remains a satirical icon par excellence of the Age of Reason. The birth of the modern world brought

forth by such intellectual giants, as Newton, Boyle, Hooke, Huygens, Leibniz, Descartes, Galileo, Harvey, Hume, Lock, Paine, Rousseau, Montesquieu, Diderot, Hobbs, Spinoza, Voltaire, Jefferson and many others, gave reason to believe that humanity was on the verge of a new age of learning where anything could be possible. Science and the new humanities could change society for the benefit of all. Science would replace religion, while the new humanism would reject the Ancien Regime and the "divine rights of kings" for the natural and inalienable rights of all humanity, radical philosophies that would manifest in the American and French revolutions.

The great "Enlightenment" was a time of supreme optimism; a word that seemed to sum up the times. And yet Francois-Marie Arouet, better known to history by his nom de plume, Voltaire, would write his most famous work as a bitter rejection of this prevailing spirit. The philosophy of scientific optimism, had it roots in the belief in the superiority of modern thought over the inherited twin intellectual authorities of the ancients and the church. No friend of the Ancien Regime, Voltaire was twice imprisoned in the Bastille, he was nonetheless no particular friend of the new "optimism" either. Against the historical backdrops of the devastating Seven Years War, a global conflict that Winston Churchill considered could have been with good reason termed the true "First World War", and the apocalyptic Lisbon earthquake of 1755 it did not seem to Voltaire that he lived in the "best of all possible worlds". Indeed this catch cry seemed little different to the sentiments of "God moving in mysterious ways".

Candide lives in Westphalia, in the splendid mansion of Baron Thunder-ten-tronckh, where he is tutored by Professor Pangloss an acolyte of the new Philosophical optimism, whose mantra he loudly proclaims to all, "we live in the best of all possible worlds", he assures Candide, and even though bad things may happen from time to time everything happens for a very good reason and so in the longer term everything always turns out for the best. But then disaster strikes when Candide is expelled from the mansion by the Baron for sexual indiscretions committed with his daughter Cunegonde. He has many horrific misadventures, characterized by exponential violence. In his travels he comes across Dr Pangloss who has also been expelled from the mansion, after the Baron had been killed by soldiers. Despite the barbarity of the wider world they witness Dr Pangloss stubbornly remains an optometrist, whist another fellow traveler they meet up with, Martin, is the exact opposite, a tiresome perennial pessimist, (a word incidentally that Voltaire appears to have invented). Later on amidst the disaster of the Lisbon earthquake they come across Cunegonde now homeless since the attack on her father and another old woman who has endured unspeakable abuses for most of her life. Candide begins to have serious doubts about the philosophy of optimism and sides with Martin the pessimist. He suddenly has to flee Lisbon for a murder he commits in self defense and so he loses Cunegonde yet again.

Eventually the party strike it rich in the land of Eldorado in South America, but anxious to find his beloved again the Candide heads the party back to Europe. He discovers that Cunegonde is now a slave in Constantinople and so off they go to the lands of the Ottomans. With the last of his Eldorado fortune he secures the freedom of Cunegonde and buys a small farm outside of Constantinople, where by hard back breaking work he and the little commune of companions he has gathered around him, over his long journeys manages to make a reasonably comfortable living for all. Cunegonde sadly has

not aged well on account of her hard life and Candide no longer desires her, but he marries he in any case, and ends up being reasonably happy for the first time in his life. In the evenings after working in the fields he sits at the dinner table arguing philosophy with Dr Pangloss and Martin. They cannot decide whether all has been for the best - and so seek out the wisest Turkish Philosopher in all of the Ottoman Empire. The philosopher is a cantankerous old bastard however, and simply slams the door in their faces! On the way back to their farm they meet a simple happy and kind old Turkish farmer who offers the weary travelers his generous hospitality, they learn that he has no philosophy on life at all, other than trying to be happy with what he has got, and minding his own business. His farm meets his needs and saves him from the evils of the world, and that is all he desires - he is the happiest man in the world! Candide decides that this is how his own little commune will find ultimate happiness

Candide is in large part autobiographical. Voltaire after publishing his supposedly seditious work "Letters Concerning the English Nation", 1734, is, like Candide, forced into exile for over a quarter of a century. He lived for years in Cirey with his married mistress Madame du Chatelet (whose husband remained cooperatively absent for most of the time), but later they tired of each other. He lived for a time at the Court of Frederick the Great, until they quarreled and he had to flee again. He became wealthy however on account of his incredibly popular seditious writings. Out of favour with two absolutist courts, he had to live next in the Swiss city state of Geneva, where he became, as did Candide, the head of a little self -sustaining commune that included his niece, where in his old age after years of exile and hardship he was finally at peace and happy. In the last year of his life he was allowed to return to Paris where he had become a stupendous celebrity and hero on account of his many campaigns against the shocking social injustices of the Ancien Regime.

So what is it about Voltaire's Candide that keeps it relevant today? The answer lies in the timeless and cross cultural messages it conveys about the human struggle to understand the Universe or perhaps even the particular reality of an infinite array of possible alternate Multiverses, we find ourselves in. We struggle with the "meaning" of our existence - do we inhabit the best of all possible worlds, as the optimist Dr. Pangloss would have it, or do we in fact inhabit the worst of all possible worlds, as the pessimist Martin would have it. Or is it simply, as Candide comes to recognize, that the world is what it is for no particular reason whatever, and so we may as well just get on and make the most of the deal we have been dealt, and simply be happy the best way that we can.

Nature not infrequently throws up deleterious anatomical aberrations which modern medical science, thanks to the great founders of the Enlightenment may be able to explain, however this is but small comfort, or indeed no comfort at all, to those so afflicted. Instead, they may agonize a far deeper age old question, "why me?" Perhaps they may take the Martin view of life and live in hourly dread of the moment that their cyst will "inevitably" bleed or burst. Or perhaps they may take the Dr Pangloss view - everything is for the best - if a large intracranial extradural bleed occurs from a head injury then perhaps some valuable extra time will be had to get to surgery on account of the extra space within their heads! But really the best view of all could be Candide's. It is very unlikely that these cysts will cause any problem at all, so just be happy, forget it and get on with life!

ARACHNOID CYST

Introduction

Arachnoid cysts are collections of CSF within the arachnoid membranes, with the CSF secreted by arachnoid cells lining the cyst.

These cysts are not uncommon, and are usually benign and asymptomatic.

They are usually discovered on CT scans as **incidental findings**.

Rarely they can cause significant complications that require intervention. These include:

- 1. Expansion with mass effect/ hydrocephalus
 - Headache, vomiting seizures, focal neurologic deficits, cognitive impairment, altered conscious state.
- 2. Rupture
- 3. Bleed into the cyst

History

The Galassi classification of middle cranial fossa arachnoid cysts is used to classify arachnoid cysts in the middle cranial fossa, which account for 50-60% of all arachnoid cysts.

Galassi et al. published this classification in 1982.

Epidemiology

The incidence of asymptomatic arachnoid cysts is increasing as more patients undergo neuroimaging procedures for unrelated symptoms

On MRIs, the prevalence of arachnoid cyst is around 1.4 %. Of these, only 5 % are symptomatic. Of those arachnoid cysts that are symptomatic, 75 % occur in children.

Pathology

Arachnoid cysts are relatively common benign and asymptomatic lesions that occur in association with the **central nervous system**.

They can occur both within the intracranial compartment (most commonly) or within the spinal canal.

They are usually located within the **subarachnoid space** and **contain CSF**.

Cause:

Arachnoid cysts are thought to arise due to the **congenital splitting of the arachnoid layer** with **accumulation of CSF within this potential space.**

The cyst wall is comprised of flattened arachnoid cells forming a thin translucent membrane. There is no solid component and no epithelial lining.

Locations:

Arachnoid cysts can occur anywhere within the central nervous system; essentially, where there is arachnoid membranes.

Most frequently (50-60 %) they located in the middle cranial fossa, where they invaginate into and widen the Sylvian fissure.

In this location they can be classified into three types based on their size according to the Galassi classification, (see Appendix 1)

Other less common locations can include:

- Suprasellar cistern (suprasellar arachnoid cyst)
 - Suprasellar cysts may cause obstructive hydrocephalus or occasionally and/or endocrine dysfunction.
- Within the ventricles (intraventricular arachnoid cyst)
- Posterior fossa
- Cisterna magna (needs to be distinguished from a mega cisterna magna)
- Cerebellopontine angle (needs to be distinguished from an epidermoid cyst)
- Spinal canal

Complications:

Rarely arachnoid cysts can cause significant complications that require intervention. These include:

1. Expansion with mass effect/ hydrocephalus

The mechanism of cyst enlargement over time is not well understood, theories include:

Passive diffusion of CSF into the cyst

- Progressive entrapment of CSF within the cyst due to a ball valve effect.
- 2. Rupture
- 3. Bleed into the cyst
- 4. Subdural hematomas may occur following relatively minor head trauma.

Clinical features

The vast majority of arachnoid cysts are **asymptomatic**.

When symptoms occur, they are usually the result of:

1. Gradual enlargement resulting in mass effect.

Features of raised intracranial pressure:

- Headache
- Vomiting
- Confusion
- Seizures
- Altered conscious state
- 2. An acute complication such as a bleed or rupture.

Investigations

CT Scan:

On CT imaging, arachnoid are characterized as follows:

- Well circumscribed lesions.
- Imperceptible walls
- Filled with CSF hypodensity fluid
- Larger lesions may show some mass effects including chronic remodeling of adjacent bone.

CT cisternography is introduction of contrast into the subarachnoid space in order to demonstrate communication of the cyst with the subarachnoid space. 3-10 mL of an

iodinated nonionic low-osmolar contrast agent is installed into thecal sac after lumbar puncture.

As this communication is slow, the cyst often fills later, and contrast may be seen to pool with it, outlining its dependent portion.

MRI Scan:

As arachnoid cysts are filled with CSF they follow CSF on all MRI sequences, including FLAIR and DWI. This enables them to be distinguished from epidermoid cysts for example.

MRI shows CSF characteristics as low density on Tl-weighted and high density on T2-weighted images and no enhancement.

As the arachnoid cyst wall is very thin it is only occasionally seen, and displacement of surrounding structures implies their presence. As there is no solid component, no enhancement can be identified.

Phase contrast imaging can also be employed not only to determine if the cyst communicates with the subarachnoid space, but also to identify the location of this communication.

Magnetic resonance cisternography is a technique that uses high resolution sequences such as CISS & FIESTA to help to delineate arachnoid cyst walls and adjacent anatomic structures.

Differential diagnosis:

General imaging differential diagnosis considerations include:

- Enlarged CSF space (e.g. mega cisterna magna)
- Epidermoid cyst
- Subdural hygroma/chronic subdural haemorrhage
- Other non-neoplastic cysts
- Neurocysticercosis
- Old infarcts
- Low-grade gliomas

Cyst fluid:

Arachnoid cysts contain clear CSF with a normal cell count and protein.

Hemorrhage into the cyst may cause xanthochromia.

A markedly elevated protein or pleocytosis suggests the possibility of cystic neoplasm rather than an arachnoid cyst.

Management

Arachnoid cysts are benign, and the vast majority remain asymptomatic throughout life.

Larger lesions may be followed with serial imaging and neurologic clinical assessments.

If symptoms occur then surgery may be warranted.

Surgical options include:

- 1. Craniotomy with fenestration into the subarachnoid space
- 2. Excision
- 3. Placement of a cysto-peritoneal shunt.
- 4. Needle aspiration:
 - This may provide temporary benefit but is not a good long-term treatment option

Appendix 1

Galassi classification of middle cranial fossa arachnoid cysts:

The **Galassi classification** of middle cranial fossa arachnoid cysts is used to classify arachnoid cysts in the middle cranial fossa, which account for 50-60% of all arachnoid cysts.

The classification uses the **size** and **degree of displacement of the adjacent brain** to divide cysts three types.

The size also correlates with the ease with which the cyst communicates with the subarachnoid space as discerned on CT cisternography or phase contrast MRI

The classification is as follows:

Type I:

- Small, spindle-shaped
- Limited to the anterior portion of the middle cranial fossa, below the sphenoid ridge
- Free communication of subarachnoid space

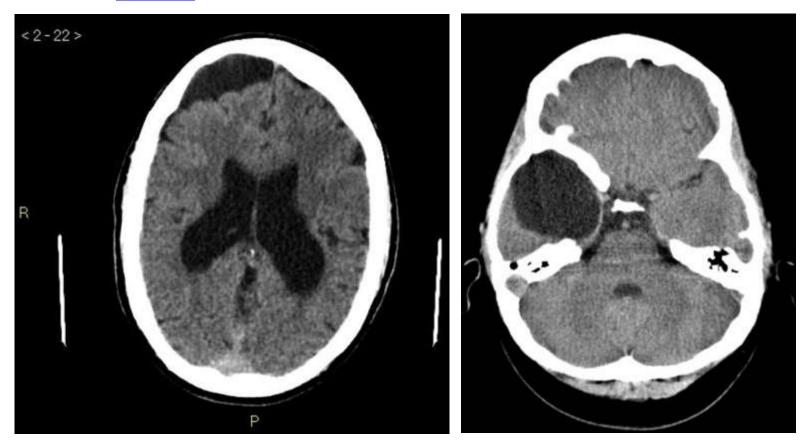
Type II:

- Superior extent along Sylvian fissure
- Displacement of the temporal lobe
- Slow communication with subarachnoid space

Type III:

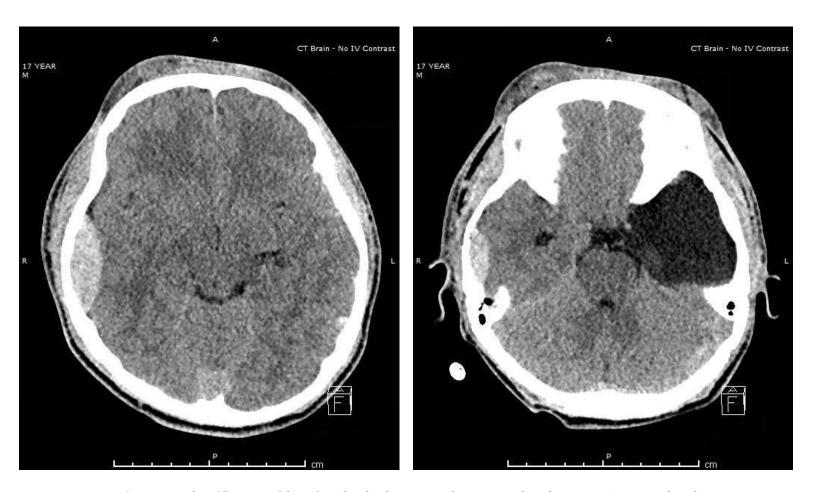
- Large, fills the whole middle cranial fossa
- Displacement of not only the temporal lobe but also the frontal and parietal lobes
- Often results in midline shift
- Little communication with subarachnoid space

Appendix 2



Left: Typical CT scan appearance of an arachnoid cyst within the tight anterior cranial fossa, (Case courtesy of Dr Sajoscha Sorrentino, Radiopaedia.org, rID: 19826).

Right: CT scan of a large Type III Arachnoid Cyst of the right middle cranial fossa, (Case courtesy of A.Prof Frank Gaillard, Radiopaedia.org, rID: 2581)



CT scan of a 17 year old male who had sustained a severe head injury. An extradural hematoma is seen on the right. A large, incidental, type III middle cranial fossa arachnoid cyst is seen on the left - this may have allowed for some fortuitous temporary pressure relief from the rapidly expanding extradural hematoma! (CT images courtesy Dr Amy McAllister).

<u>References</u>

- 1. Milan G Chheda et al. Arachnoid cyst in Up to Date Website, December 18, 2017.
- 2. Yuranga Weerakkody, Frank Gaillard et al. Arachnoid cyst in Radiopedia, Website:
 - https://radiopaedia.org/

Dr. J. Hayes March 2018