CASE REPORT

Spinal meningioma, aortic aneurysms and the missing link of observation: the anchoring heuristic approach

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SUMMARY

Over the last three decades, the development of systematic and protocol-based algorithms, and advances in available diagnostic tests have become the indispensable parts of practising medicine. Naturally, despite the implementation of meticulous protocols involving diagnostic tests or even trials of empirical therapies, the cause of one's symptoms may still not be obvious. We herein report a case of chronic back pain, which took about 5 years to get accurately diagnosed. The case challenges the diagnostic assumptions and sets ground of discussion for the diagnostic reasoning pitfalls and heuristic biases that mislead the caring physicians and cost years of low quality of life to our patient. This case serves as an example of how anchoring heuristics can interfere in the diagnostic process of a complex and rare entity when combined with a concurrent potentially life-threatening condition.

BACKGROUND

Over the last three decades, the development of systematic and protocol-based algorithms and advances in available diagnostic tests have become the indispensable parts of practising medicine. The ease of acquiring advanced imaging to rule in or out a life-threatening diagnosis, irrespective of the clinical question's pretest probability, ironically both saved lives and generated issues of inappropriate test utilisation. Naturally, despite the implementation of meticulous protocols involving diagnostic tests or even trials of empirical therapies, the cause of one's symptoms may still not be obvious; therefore, management at that point necessitates a reset and a return to an individualised diagnostic approach. In the setting of a symptom refractory to standard diagnostics and trial therapy, the physician is left uncertain as to whether the correct diagnosis has not yet been discerned, or if the correct diagnosis is made but provided treatment is inadequate.

Here, we report a case of chronic back pain, which took about 5 years to get accurately diagnosed. The case challenges the diagnostic assumptions and sets ground of discussion for the diagnostic reasoning pitfalls and heuristic biases that mislead the caring physicians and cost years of low quality of life to our patient.

CASE PRESENTATION

A 61-year-old woman was diagnosed with Stanford type A aortic dissection (AD) in 2006 after complaining of back pain in the thoracic region. At

that time, she underwent surgical repair of the proximal ascending aorta. One year later, a dissection to the descending thoracic aorta starting at the level of the left subclavian artery was seen by follow-up CT angiography (CTA). The lumen expansion was less than 50 mm and annual follow-ups were recommended. Surgical correction was pursued again 3 years later once lumen reached diameter of 66 mm due to the high risk of rupture.

Meanwhile, the patient continued complaining of back pain in the thoracic area, associated with physical activity. As part of the workup of her symptoms, she underwent a nuclear stress test, which was inconclusive for anterior wall ischaemia and subsequently a coronary angiography, which did not show evidence of coronary disease. Eight months after the last CTA in 2009, she underwent a second vascular reconstruction with a thoracic aorta endovascular reconstruction after a left subclavian artery to left common carotid artery transposition. Three months after the reconstruction, she was seen in the clinic and her back pain was still present but milder.

About 5 years later, in January 2015, her back pain was again severe enough to send her to the emergency department. She was found to have elevated troponins and a diagnosis of a non-ST-elevation myocardial infarction was made. The coronary angiography again failed to show significant coronary artery disease. Because of her previous history, a CTA was performed showing an expansion of the thoracic descending aorta beyond the old stent graft. She underwent stent graft placement in the thoracic aorta but the back pain was not resolved this time. Three months after her last surgery, she was readmitted with acute back pain. The CTA showed no signs of aortic rupture or a new dissection but it demonstrated a mass in the spinal cord (figure 1). A magnetic resonance tomography (MRT) revealed a meningioma of the spinal cord, at the level of the sixth thoracic vertebral body, and she was referred to the neurosurgical department for a microsurgical removal of the mass. After the tumour resection, she was completely asymptomatic.

The first neurological examination was documented in 2011 and the only pathological findings found were hypaesthesia of the lateral margin of the left foot and bimalleolar pall-hypoesthesia 5/8. There were no pathological findings in the neurological examination of the lower limbs or sphincter functionality. The neurological examination in the neurosurgical department in 2015, after the diagnosis was set, revealed no sensomotoric deficits of the extremities, bladder or sphincter dysfunction.

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Figure 1 CT angiography demonstrating the progressing spinal meningioma size. Above: December 2006. Middle: December 2012. Bottom: October 2015 (arrows indicate the meningioma; lines show the aneurysm size).

The patient had dorsalgia over the left paraspinal region with increase in stress. In every neurological examination throughout the years, no lower limbs or sphincter dysfunction was documented.

OUTCOME AND FOLLOW-UP

For about one decade, this patient underwent more than 13 CTAs, 3 MRTs, 2 coronary angiographies and presented to the emergency department more than 30 times. During this time, the

Figure 2 Magnetic resonance tomography (axial and sagittal views), performed in October 2015, demonstrating a mass in the spinal cord (arrows) without signs of aortic rupture or a new dissection.

aortic aneurysm, the dissection or other cardiovascular diseases were entertained as the cause of her symptom. A retrospective review of the images from all the imaging performed showed that the spinal mass could be seen in 2006. A slow but constant progression especially over the last few years was also evident (figure 2). The spinal meningioma became symptomatic at least the last 5 years as it was gradually enlarging since then. However, all the diagnostic efforts focused solely around cardiovascular causes.

DISCUSSION

Chronic back pain is the second most common cause of disability in US adults, one of the most increasingly care-seeking conditions worldwide and among the top 10 diseases and injuries that account for the highest number of disability-adjusted life years worldwide based on the 2010 Global Burden of Disease Study.¹ European guidelines (2004) do not recommend radiographic imaging unless a specific cause is strongly suspected.² Once imaging is obtained though, management decisions depend on the anatomical problem detected and if so usually on its evolution over time.

Spinal meningiomas on the other hand constitute an understudied medical entity. Many meningiomas are diagnosed incidentally with imaging obtained for the evaluation of another condition. Only recently, Kshetry *et al* provided epidemiological data for the disease, with the US incidence being estimated at 0.33/100,000.³ The majority of spinal meningiomas are benign (WHO grade I) but a small percentage are atypical (WHO grade II) or anaplastic (WHO grade III).⁴ The disease is rare, and it may have a long latency of 20–30 years or more. The recent advances in neuroimaging and the magnetic tomography increased the incidence of meningiomas in the post-CT decade. Therefore, the prevalence of subclinical disease is assumed to be higher than those with clinically confirmed diagnosis. Depending on the location of the tumour and the pressure of the growing tumour on the spinal cord or spinal nerves, spinal meningiomas may cause motor deficits like weakness or numbness and less frequently sensory loss, pain, bladder, bowel and/or sexual dysfunction. Male sex and tumours with radiological evidence of a dural tail are proven to be associated with an increased risk of recurrence after surgical treatment.⁵

Haemodynamic changes are associated with rapid tumour progression. Meningiomas in pregnancy show also fast progression, due to the hypervascular state.⁶ In our case, the back pain

recurred after overstenting the spinal arteries. Following that procedure, the spinal cord blood supply demand and the development of new collateral arteries could probably explain the growth of the tumour and the progression of her back pain. This could explain the recurrence of the back pain after the stenting.

The incidence of AD is estimated at six per hundred thousand persons per year.⁷ Abrupt onset of severe chest and/or back pain is the most common clinical presentation. Surgery is the treatment of choice in type A AD, because the mortality can reach 50% within the first 48 hours if not operated. Early detection with imaging guides the surgical intervention of an otherwise fatal condition.

When a non-specific symptom is coupled with another coexistent potentially life-threatening disorder, the diagnostic clarity is nuanced. The goal of the present case is to identify and discuss anchoring heuristics, to outline the risk factors to develop ones and explore ways to minimise and identify such errors. By no means is exhaustive but it is a representative specimen of daily practice pitfalls and highlights the importance of acquiring an integrated approach towards the clinical question before proceeding in unnecessary and frequently unpleasant investigations.

Schiff *et al* reported that about 75% of diagnostic errors have a cognitive component with 32% of error being attributed to clinician assessment errors (failure to consider and overweighing competing diagnosis).⁸ Other studies show that most errors will involve common diseases with atypical presentations or failure to consider alternative diagnosis.⁹ Rarer diseases can be even more difficult to detect, especially when the presenting symptoms are non-specific.

Extensive cognitive psychology research over the years explains how physicians process information and develop medical plans. As applied to healthcare, heuristics are shortcut mental strategies that allow faster processing of information but can lead to error because of the exclusion of important information.¹⁰ Biases are faulty beliefs that affect decision-making.¹¹

The theory of heuristics suggests that clinicians lock onto an initial diagnostic hypothesis (an anchor) prematurely and they fail to adjust this hypothesis in light of later information. Anchoring exacerbates premature closure; the failure to consider alternative diagnoses after the initial impression is formed. The clinician's attention is focused solely on information that supports the initial hypothesis and data that are inconsistent with this hypothesis are often being disregarded. In this case in particular, anchoring appears to have been strong, where despite the persistent symptoms the management plan was simply repeated for half a decade.

A series of cognitive biases contribute to anchoring. The anchoring bias is only one contributor towards the overall tendency to anchoring and premature closure. Factors that trigger awareness to certain cues and not to others have not been identified. The effects of environmental elements such as stress or time constraints also play a role. The tendency to selectively seek information supporting the initial diagnosis, confirmation bias, is an important factor in that context. The most effective way to reduce this bias is to always seek information that has the potential to lead away the current impression. Some physicians exhibit the tendency to overvalue information irrelevant to the case if it has been discovered purposefully by the clinician. The latter often contributes to the confirmation bias since the clinicians first seek and overvalue irrelevant information when obtained.

Another cognitive bias contributing to anchoring is the status quo bias, where the clinician tends to insist on the initial impression as the number of alternative diagnoses increases. The more complex and broader differential paradoxically may increase the

clinician's anchoring. Lastly, framing can influence the clinical judgement when forming a diagnostic impression. Our patient was given the label of a 'vascular' patient enhancing the anchoring. Dedicated specialists have had their brain conditioned to work in the differential diagnostic frames within their specialty.

Cognitive awareness and system-based practices can reduce diagnostic errors. Simply teamwork and case discussions can increase cognitive awareness through self-reflection and thought process enhancement. Structured diagnostic protocols of common clinical scenarios in any form offer guidance on decision-making to clinicians improving diagnostic accuracy.

Besides the reasoning process, a missed diagnosis may be related to diagnostic tests done, to diagnostic tests not done, diagnostic tests that were faulty translated or diagnostic results that were not followed up. Nine years after the initial diagnosis of an aortic aneurysm was made, more than twelve CTAs and three surgeries later, the patient had persistent back pain. Interestingly, the spinal meningioma was present even in the initial imaging studies, but its presence was never perceived as 'actionable' or 'in the context interpretable' information' to her physicians' until years later.

Medical technologies have improved the diagnostic accuracy and therefore the treatment processes, but they also come with an increase in healthcare costs. The availability of technologies like CT scanners and MRI units has increased over the last two decades in the Organization for Economic Co-operation and Development (OECD)-participating countries. Although in Germany the use of these tests is clearly lower than the average of the OECD countries, examples such as our patient alert us to stay vigilant.

Guidelines and campaigns have been developed in several countries such as UK (National Institute for Health and Clinical Excellence) and the USA (Choosing Wisely, American College of Radiology Appropriateness Criteria) to limit the excessive use of them.

Learning points

- ▶ This case serves as an example of how anchoring heuristics can interfere in the diagnostic process of a complex and rare entity when combined with a concurrent potentially life-threatening condition.
- ▶ Recognising the potential of these biases is an integral part of systems based practice and is absolutely necessary for the accreditation and credentialing of physicians.
- ▶ As more literature arises, we may be able to determine the circumstances predisposing healthcare workers in making these errors leading to either diagnostic errors or delayed diagnosis.
- ▶ In the meantime, self-awareness and self-reflect on the above, along with multidisciplinary approach of a problem, will guide physicians towards the right diagnosis.

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