An unusual cause of ear pain. A life threatening disease revealed by a common symptom

耳痛的不尋常原因。一個常見症狀揭示的危及生命的疾病

M Sierecki, M Marchetti, M Verdier, A Ghazali

A 40-year-old woman presented to the emergency department (ED) for bilateral otalgia as her sole complaint. The physician's otoscopic examination was normal and the rest of the examination was unremarkable except for previously unknown high blood pressure. The patient had no chest pain or dyspnea. She was discharged from the ED with antihypertensive therapy, pain-relief medications, and an appointment with a cardiologist. Twelve days later, transthoracic echocardiography revealed pericardial effusion and dilated ascending aorta. Computed tomography scan finally diagnosed an aortic dissection (AD) type A (Stanford classification) which necessitated emergency surgery. (Hong Kong j.emerg.med. 2016;23:376-379)

一名 40 歲的女性到診急診科（ED），主訴為雙側耳痛。耳鏡檢查正常，除了先前未知的高血壓，其餘的檢查無明顯的異常。患者沒有胸痛或呼吸困難。患者從ED出院，醫生開了抗高血壓以及緩解疼痛的藥物，並安排預約心臟專家會診。12天後，經胸超聲心動圖顯示心包積液和升主動脈擴張。CT掃描終於確診 A 型（斯坦福分類）主動脈夾層（AD），需要進行急診手術。

Keywords: Aortic dissection, humans, referred pain

關鍵詞：主動脈夾層、人類、轉移痛

Case presentation

A 40-year-old woman presented to the emergency department for a sudden bilateral ear pain that had occurred the day before. She had a known history of migraine and was treated by Propranolol 40 mg. Her cardiovascular risk factors included obesity and family history of coronary disease and high blood pressure. She was independent and worked in a printing house. Clinical exam was performed by a senior physician. Otoscopic and oral cavity examination was unremarkable. There was no complaint of headache and no symptoms of meningitis. The patient had no fever. The heart and lung auscultation was unremarkable. She had neither chest pain nor shortness of breath. On admission, vital signs were: heart rate was 86 bpm, blood pressure (BP) was 230/140 mmHg, temperature was 37°C, SpO_2 was 99% on room air. The blood sugar level was 1.16 g/L.

Electrocardiogram (ECG) and blood tests were ordered. Initial ECG showed no repolarisation abnormality and no QRS microvoltage. To rule out

Correspondence to:
Mathias Sierecki, MD
University Hospital Center of Poitiers, Department of Emergency Medicine, 2, rue de la Milétrie 86000 Poitiers, France
Email: math.sierecki@gmail.com

Matthieu Marchetti, MD
Aiham Ghazali, MD

University Hospital Center of Poitiers, Department of Radiology, 2, rue de la Milétrie 86000 Poitiers, France
Marine Verdier, MD

nephritic syndrome, a urine dipstick was done and showed blood 1+ and no protein. Blood test showed no renal failure (creatinine=84 micromoles/L) and normal electrolytes (K=3.6 mmol/L). Another blood test was prescribed for the day after. It showed reduced renal function (Creatinine=110 micromoles/L, Glomerular filtration=48 mL/min/1.73 m^2 MDRD), a Haemoglobin level at 14.1 g/dL, 19,700/mm^3 leucocytes and C Reactive Protein=232 mg/L. TSH was normal (3.1 mUI/L).

The physician focused on the high BP and made no link between ear pain and high BP, judging by good clinical tolerance and normal investigations. The patient was discharged from ED with antihypertensive medications (amlodipine and valsartan) and an appointment with a cardiologist. Furthermore, she was prescribed additional blood tests (electrolytes, TSH, creatinine, CRP and complete blood count) and ambulatory blood pressure monitoring. The final diagnosis for her ear pain was angina tonsillitis with referred pain. When the patient presented to her consultation 12 days later, transthoracic echocardiography (TTE) revealed circumferential pericardial effusion, ascending aorta dilatation, concentric left ventricular hypertrophy with preserved global left ventricular function, and no aortic regurgitation. These findings led to an urgent CT scan that finally diagnosed an aortic dissection (AD) type A of Stanford. It was extended from aortic root to brachiocephalic trunk without dissection of coronary arteries (Figure 1). It was complicated with a false aneurysm of the aortic arch of 49 x 19 x 47 millimeters (Figure 2).

The patient received open surgical repair with replacement of ascending aorta under hypothermic circulatory arrest. She experienced acute ischaemia of the right arm with rhabdomyolysis and acute renal failure necessitating surgical fasciotomy and 5 sessions of dialyses. After 17 days in intensive care unit and 4 months of hospitalisation, the patient was discharged home with no persistant ear pain. She had 50% handicap of the right arm despite occupational therapy. Yearly follow-up consultation was planned with cardiothoracic surgeons and cardiologists.

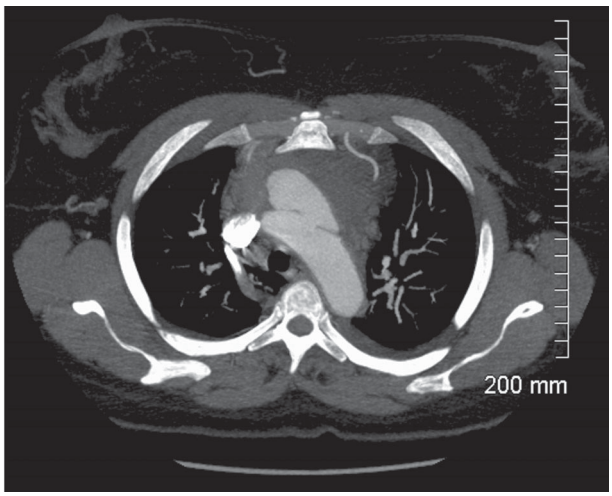

Figure 1. Transversal computed tomography scan view of dissection Stanford type A from aortic root to innominate artery.

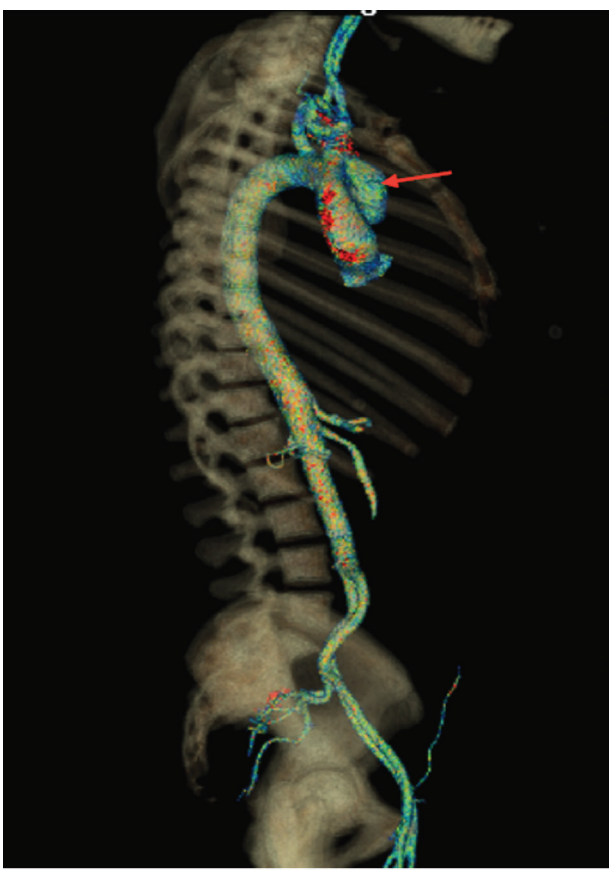

Figure 2. 3D reconstruction view of false aneurysm of the aortic arch (arrow) of 49 x 19 x 47 millimeters.

Discussion

Surgical repair with replacement of ascending aorta under hypothermic circulatory arrest was regarded as the gold standard procedure.[1] Despite improved therapeutic techniques per operatory mortality is as high as 5% and follow-up mortality is up to 35% within 30 days.[2] Bleeding, ischaemic complications (stroke, mesenteric infarction) and wound complications (dehiscence, infections) were most frequently described. Cardiac side effects of this surgery included myocardial infarction, tamponade or low cardiac output syndrome.[2]

To our knowledge, only two cases of ear pain as the sole symptom of AD had reported in literature.[3,4] In both cases, the patient died before surgery. The discovery of high BP, which was the most prevalent risk factor (75%),[5] and the sudden onset of pain[6] should had alerted the physician. But the lack of chest pain, the good tolerance of high BP, and the atypical association of symptoms erroneously reassured the physician.

There are two hypotheses to account for the ear pain in AD. The ear receives cutaneous innervation from four cranial nerves:[7] trigeminal (V), facial (VII), glossopharyngeal (IX), and vagus (X); and two spinal nerves from the upper cervical plexus (C2, C3). The aortic and cardiac extrinsic innervation is supplied by cardiac and pre-aortic plexus where various vagal and sympathetic nerves end. The parasympathetic innervation is supplied by three main nerves: the superior and inferior cervical cardiac branch of the X mainly contributing to anterior cardiac plexus, and the thoracic branch of the X. They originate in the inferior vagal ganglia. The parasympathetic system carries signals under the control of higher centres in the in medulla oblongata.[8]

In cases of myocardial infarction, several authors have suggested the role of the auricular branch of vagus nerve[9,10] (also called Alderman's nerve) which innervates the concha and parts of the external auditory meatus.[7] Chen at al explained this phenomenon as a "sensory error".[11] Indeed the convergence of the complex sensory innervation for both the ear and cranial nerves by common sensory pathways sometimes renders the central nervous system unable to exactly pinpoint the origin of pain. This mechanism could explain exclusive craniofacial pain in 6% of acute coronary syndrome.[12,13] These neuroanatomic findings could explain the occurrence of ear pain during AD in our case.

The mandibular branch of trigeminal nerve (V3) supplies sensory innervation of the pinna, the tragus, the anterior and superior walls of the external auditory canal, and the surface of the tympanic membrane.[14] Several studies have reported pain in regions innervated by the trigeminal nerve following electrical stimulations of the vagus nerve.[15-17] This suggested relationships between these two pathways and could explain the occurrence of otalgia in acute coronary syndrome for.[18,19] Chandler's research on rodents[20] and primates[21,22] found that electrical and chemical stimulations of left vagus nerve caused activation of C1-C2 left spinothalamic tract neurons at the level of the trigemino-thalamic tract. Because sensory innervation of the ear is also supplied by spinal nerves from the high cervical plexus, it offers a possible physiologic explanation for otalgia in thoracic pathology especially AD. Further research is needed.

This case report highlights the fact that a very common symptom can reveal a deadly disease. Considering the individual variation of anatomy and the rareness of such cases (due to high mortality rate for missed diagnoses), specific studies of otalgia in AD are not feasible.[23] Even so, the knowledge of the neural pathways contributes to understanding of such mechanisms of pain and physician might use simple investigations to decrease the risk of misdiagnosis. First of all, BP is routinely measured in all patients presenting to ED and should be done on both arms in any case of high BP with sudden onset of pain. Moreover, this case suggests that every case of sudden onset of bilateral otalgia with a normal otoscopic and oral cavity examination might require ECG and chest X-ray (CXR) evaluation. This might rule out a cardiac cause especially in patients with high BP. CXR has a low sensitivity (67% for AD[24]) but the discovery of a widened mediastinum[25]

might warn the physician of the need to perform tomographic aortic imaging. In this case, no CXR was performed and missed the chance to make a diagnosis earlier. In general, physicians should keep in mind that half of otalgias are referred pain due to irritative lesion involving the fifth, ninth, or tenth cranial nerves and spinal nerves C2 and C3 It may be an early harbinger of serious underlying pathology. Physician should be vigilant in cases of bilateral pain with no accompanying otologic symptoms.[7,11,26,27]

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