Case report: dysphagia and sleep apnoea associated with cervical osteophytes due to diffuse idiopathic skeletal hyperostosis (DISH)

Diffuse idiopathic skeletal hyperostosis (DISH) may be an under recognised contributory factor to both dysphagia and obstructive sleep apnoea.

A seventy-two year old man presented with a 12 month history of intermittent painless dysphagia. He had been aware of the need to be careful when eating and coughing after swallowing was a daily occurrence. Liquids were more troublesome than solids. He had not lost any weight.

In 1989 he had been diagnosed as having obstructive sleep apnoea syndrome. Over-night impedance pneumography revealed more than 40 episodes of apnoea per hour and simultaneous oximetry showed a corresponding fall in oxygen saturation to 80%. Treatment with nocturnal continuous positive airway pressure (CPAP) resulted in improved sleep quality and daytime wakefulness. There was a history of ischaemic heart disease and atrial fibrillation.

On examination he was 170 cm tall, weighed 87 kg, and his neck circumference was 46 cm. He was in controlled atrial fibrillation. There were no neurological signs but the tongue was large and the oropharynx appeared small. Cervical spine movements were limited in all directions. In a timed test of swallowing he coughed violently after swallowing the first bolus. After a pause he was able to drink 120 ml in 50 seconds (2.4 ml s⁻¹; normal >10 ml s⁻¹). Over a two week period his swallow speed fluctuated between 6 ml s⁻¹ and 20 ml s⁻¹; a double blind edrophonium test had no effect on swallowing speed.

Videofluoroscopy examination showed partial hold up of the bolus of contrast in the mid-cervical region. Lateral cervical spine radiographs showed a bridging mass of exuberant new bone formation anteriorly extending from C2 to C7, with a dividing pseudoartefact at the C3/C4 level (figure). A bony canal stenosis was also present from the C2 level downwards. Radiographs of the thoracolumbar spine showed areas of new bone formation bridging the vertebral bodies anteriorly. Despite the extent of the new bone formation the intervertebral disc spaces were relatively well preserved. Radiographs of the pelvis showed irregular projections of bone from the iliac crests and the lateral margins of the ileum. The radiographic features were typical of DISH. An MRI scan of the brain stem and a CT scan of the brain were both normal. A 75 g glucose tolerance test with capillary blood suggested diabetes mellitus with a fasting value of 7.1 mmol/l and a 120 minute value of 11.2 mmol/l.

The patient received advice from the speech therapist and was able to reduce the frequency of coughing by taking double swallows for each bolus and slowing down his eating. He did not wish to be considered for any type of surgical intervention and continues on CPAP at home. Follow up for three years has revealed no other disorder.

In this case there is good radiographic evidence to suggest that the limitation of neck movement and the large anterior cervical osteophytes were due to diffuse idiopathic skeletal hyperostosis (DISH). Forestier’s original description of the condition was confined to the spine but many extraosseous manifestations have now been described. The condition is common, occurring in 6%-12% of a necropsy population, and impaired glucose tolerance is often a feature.

Although the patient had other features that predisposed him to develop the sleep apnoea syndrome including a large tongue and neck circumference, the C2/C3 level is a common site of obstruction in this condition. It seems highly likely that the bony mass contributed to occlusion of the oropharynx during sleep and videofluoroscopy suggests that it contributed significantly to the dysphagia; barium swallow examination, indirect laryngoscopy, and neurological investigation revealed no other cause for the dysphagia.

Dysphagia is a recognised symptom of cervical spine involvement in patients with DISH. Obstructive sleep apnoea is an association not described before although cases of stridor due to impingement of the osteophyte on the laryngeal vestibule have been reported.

The case shows how a lateral cervical spine radiograph may be helpful in the diagnosis of patients with dysphagia and/or sleep apnoea without neurological signs. In view of the frequency of anterior cervical osteophytes in the older population it is imperative that other causes be excluded; oesophageal perforation after endoscopy in patients with large cervical osteophytes has been reported and barium examination of the oesophagus may be preferable.

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Horologogia: an impairment of the ability to tell the time

We report a 57-year-old male patient who had a right posterior stroke. A CT scan indicated extensive hypodensity within the territory of the right posterior cerebral artery. There were also lacunes in the lateral thalamus and corpus striatum bilaterally, as well as small scattered lesions, predominantly within the peripheral areas of white matter of the posterior frontal and parietal lobes, consistent with ischaemic changes due to small vessel disease. On admission to hospital he presented with decreased sensation and muscle weakness on the left side of his body, left homonymous hemianopia and left-sided visuospatial neglect. Neuro-psychological assessments during the first three weeks after his admission to the hospital showed well preserved language, verbal reasoning, and verbal memory skills (average range or above). On a relatively easy picture recognition memory test he also performed satisfactorily. Left-sided visuospatial neglect was evident in his performance on line bisection, cancellation, and search tasks. He easily identified real objects and line drawings of objects or
photographs of certain famous faces. His performance on the fragmented letters test of visual perception was, however, impaired. On the cube analysis test of visuospatial analysis his performance suggested that he was able to integrate stimuli (cubes) into spatial (three dimensional) schemas but he obtained a low score apparently due to the high number of expressions which were negated in his responses. He was able to draw a cube (without a model), making only slight mislocations of certain lines depicting depth.

From the time of his admission to the hospital, he had been presented with a profound difficulty in telling the time from a clock (or a watch), even when the clock showed a simple "on the hour" time. We investigated this impairment in two experimental sessions, two weeks and four weeks after his admission. In each session the following four conditions were used, involving "time telling or clock setting for eight different "on the hour" times, chosen to represent equally the numbers on the left and right hand side of the clock. Each of eight "on the hour" times was presented twice in a random order. (1) The patient was shown a cube face with a projected time (12:05 cm) indicating an "on the hour" time. He was asked first to name the time, then to report the position of the clock hands and, finally, to tell the time depicted on the clock (correct scores were scored: correct if he gave the appropriate answer before or after identifying the positions of the clock hands; his scores in the two sessions were 0/16 and 0/16 (correct). (2) He was shown a drawn clock face minus the hands on which he was required to insert the hands to depict a verbally given on the hour time; he scored 16/16 (correct) in both sessions. (3) The positions of the hands for an on the hour time were described verbally to the patient who was then required to say what that time was; his scores in the two sessions were 4/16 and 6/16 (correct). (4) He was given the time verbally and was then asked to say in which positions the hands should be; his scores were: 16/16 and 15/16 (correct). The results from conditions (2) and (4) clearly indicate a dissociation between his virtually errorless performance in conditions (2) and (4) where the response involved clock setting and his very poor performance in conditions (1) where involved time telling from the clock face. His deficit in condition (1) occurred despite consistently accurate and unhesitating identification of the positions of the hands. When he was shown a clock face and asked to name the time, he seemed rather bewildered and reluctant to respond to the question. With encouragement he would refer to the "12" to which the long hand was pointing as "the hour and the number to which the short hand was pointing as referring to the number of minutes to, or past 12 o'clock. For example, he would refer to "eight o'clock" as "eight minutes to 12" or "eight minutes past 12" and to "four o'clock" as "four minutes to 12" or "four minutes past 12". In condition (3), in which he was asked to tell the time on a clock face, his performance was better than in condition (1) but still impaired. By contrast, in conditions (2) and (4) he would respond readily and accurately to the clock or giving verbally the positions of the short and long hands in the appropriate positions.

Three weeks after the second experimental session his performances in conditions (2), (3), and (4) were virtually perfect and half of his responses in condition (1) were correct. Deficits in time telling from a clock face have previously been reported in association with visuospatial neglect and it would be reasonable to argue that inattention to the left-hand-side of the clock underlay our patient's inability to recognize the time on a clock face. Against such an explanation would be the fact that consistently, readily, and without prompting he located accurately the left-hand-side of the clock. He was also able to set the time on demand normally, a task that necessitates accurate search of the left-hand-side. Indeed, his satisfactory performance on other tests that are dependent on accurate recognition of (non-degraded) visual stimuli suggested that his visuospatial neglect was task specific—that is, it was mainly manifested when stimuli were meaning less (for example, cancellation tests) rather than when stimuli were continuous or meaningfully integrated (for example, pictorial material depicting animals, objects, and, presumably, spatial relations). A particularly striking finding was that he could not identify the time from a clock face on which he himself had only just correctly inserted the hands to depict a specified time. This is akin to certain cases of visual agnosia and apparently counter-intuitive neuropsychological syndromes involving other kinds of symbolic activity, such as dyslexia with dysgraphia and selective impairment of auditory comprehension of words in the presence of preserved ability to produce them on demand, or vice versa. Our data suggest that in the "telling the time" task our patient was unable to access factual knowledge about clocks and time, despite the fact that he had available to him all the relevant information. Exploration of his ability to represent visuospatial relations interactively was not possible within the time before his recovery. It is, however, relevant to add that at the time of the first set of experiments he was able to navigate a car through the streets of North London. This finding would normally contradict a significant distortion of visuospatial representations.

As far as we know, this form of transient specific time telling deficit, in the context of accessible information about the positions of the hands on a clock face and intact ability to set the time on demand, has not previously been reported. We would like to suggest that this deficit be referred to as "horologagnosia" (from the Greek meaning an impairment of the ability to tell the time).

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The coin-in-the-hand test: a new "bedside" test for the detection of malingering in patients with suspected memory disorder

The detection of malingering is an important area of clinical neuropsychological practice, especially in medicolegal settings where it is often necessary to exclude simulation or exaggeration of poor performance. A number of tests have been used by clinicians and researchers to examine the issue of assessment of malingering during memory functioning, including the Rey 15-item visual memory test and the symptom validity test.

It is likely that no single test in itself will be sufficient to prove simulation of amnesia and other memory disorders, and there remains a critical need for further tests to be developed, especially those that are simple and brief to administer and that have relatively high face validity. I therefore sought to provide a new test for malingering based on the premise that if a person is able to reproduce a series of spatial patterns on the Rey 15-item test, they should still give impaired performance in some neurological patients with genuine severe memory disorder.

Two suspected malingering patients were included, both of whom were taking part in medicolegal proceedings and were suspected of simulating poor memory functioning. An amnesic group of five patients was also examined, all of whom had had herpes simplex encephalitis, and were left with dense amnesia.

The suspected malingers showed variable, usually low, test scores on general neuropsychological tests, and it was both their clinical presentation and anomalies in test scores that prompted the referral of the patient for this study. As they both performed at chance level on the test of malingering described later, the reliability of their other neuropsychological test scores must be put in question. For the purposes of this paper, therefore, I have simply presented a brief clinical profile for each case, and the reasons why each case was included in the study.

Case 1 was a right-handed woman in her 40s, who had been working in a professional job. She was involved in an accident, and was claiming compensation for significant psychological and memory difficulties that she considered were present after the accident. She had patchy memory loss for events around the time of the accident, and it was uncertain if this reflected a period of transient amnesia or if it was due to a shocked state that resulted from the accident. She indicated that her memory for the day of the event was clearer than her memory for the three to four days before the accident. A skull radiograph was reported as normal, a CNS examination showed no...
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