CASE REPORT

Two women with recurrent falls: La maladie des genoux bleus alias cryptogenic drop attacks

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Accepted 6 June 2014

SUMMARY

Two women, aged 79 and 56, reported recurrent falls for several years without any prodromes or residues, without any known features of either syncope or seizure, without any detectable causative condition and seemingly without revealing any pathognomonic signs in the clinical examination. In both cases, the falls occurred only while walking and without loss of consciousness. These falls resulted always in dropping forward with pouncing on the knees. There had never been an injury other than contusions of the knees. Indeed, inspection of the knees of one of these patients showed signs of contusion with erosions and scars of different ages, whereas on the whole body no other wounds or dermatological lesions could be detected. What could that be? Cryptogenic drop attacks—or in French and more descriptively: La maladie des genoux bleues!

BACKGROUND

Assessing patients after falling is a common task of internists, neurologists and general practitioners in the emergency room as well as in the outpatient office.

Often, the workup reveals an ordinary fall, a syncope, a prolonged collapse due to a systemic disturbance or, less frequently, a seizure. In other cases, it is at least a predisposing factor that can be identified.

Rare are pure drop attacks as we will describe here. Drop attacks are infrequent disorders with a limited differential diagnosis. We present two cases of patients with characteristic symptoms of so-called cryptogenic drop attacks. As identification largely depends on the knowledge of the typical presentation and as literature thereof is scarce, we want to recapitulate this entity with two picture book examples.

CASE PRESENTATION**Case 1**

A 79-year-old woman was sent for evaluation in June 2010 by her son, who was more concerned than the patient herself. She had suffered from four falls within the past 12 months prior to presentation.

Falling happened usually while walking and only once while standing, but never in a sitting position. She mentioned suddenly being aware of lying or kneeling on the ground. There were no prodromal signs, neither dizziness, vertigo, nausea, sweating, weakness, blurred vision, hearing loss or paraesthesia indicating an orthostatic or vasovagal syncope, nor palpitations, chest pain or dyspnoea indicating a syncope of cardiac origin. She never remembered stumbling before falling. She never got hurt

anywhere else than on her knees. Notably she never hit her head in what otherwise could have been the reason for amnesia.

Although a friend of hers witnessed one of these falls, she was not sure whether or not there had been loss of consciousness. If so, it would have been only for a part of a second. The patient herself could not tell either because it all happened too fast.

The remainder of her history consisted of well-controlled arterial hypertension, compensated hypothyroidism, degenerative changes of her thoracic and lumbar spine and beginning macula degeneration. A few days previously, she had had colonoscopy performed for intermittent diarrhoea. While biopsy results were pending, empiric treatment for mild colitis was initiated.

Her medication consisted of perindopril 5 mg, indapamide 1.25 mg, atorvastatin 20 mg, levothyroxine 0.1 mg, mesalazine 3 g, magnesium aspartate 5 mg, paracetamol 1 g and a ginkgo biloba extract. She took all of these in the morning with the exception of the magnesium preparation, which she took in the evening.

Case 2

A 56-year-old woman, an employee in a supermarket, reported in May 2012 about recurrent falls for 4 years. She mentioned falling always on her knees, and this occurred roughly 2–8 times a year at irregular intervals.

These falls happened only while walking, without stumbling, and the patient was used to wearing flat shoes. There had never been any prodromal perceptions.

She was quite sure that she had never lost consciousness. There remains a slight uncertainty about the question of loss of consciousness in some episodes, because the falls tended to occur too fast to tell exactly. However, there had never been a period of lying on the ground.

She rather felt embarrassed about falling on her knees in public and tried to walk on as normally as possible without delay, which was usually possible. Many of these falls were witnessed. Embarrassment actually seemed to be her biggest concern next to the deterioration of many pairs of trousers at the level of the knees. She had never suffered from severe injuries other than contusion of her knees.

Her medical history was remarkable for well-controlled arterial hypertension, compensated Graves' disease being diagnosed 3 years previously, mild depression and mild obesity. Her medication consisted of candesartan 4 mg and carbimazole 5–10 mg daily.

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To cite: Butsch R,
Schneemann M. *BMJ Case
Rep* Published online:
[please include Day Month
Year] doi:10.1136/bcr-2013-
200855

INVESTIGATIONS

Case 1

After falling for the first or second time, the patient was examined by another doctor and CT of the chest and abdomen with CT angiography of the pulmonary vessels as well as coronary angiography was performed. Both examinations did not reveal any pathological findings.

Clinical examination including a full neurological examination was remarkable only for a gentle systolic heart bruit over the aortic valve region without radiation and mild ankle oedema.

Haemoglobin level, sodium, potassium and calcium as well as thyroid-stimulating hormone (TSH), vitamin B₁₂ and folic acid were in the normal range. Creatinine level of 92 µmol/L indicated mild renal insufficiency. Urinalysis was normal.

Chest X-ray, ECG, Holter ECG, orthostatic test, carotid sinus massage, echocardiography, EEG and MRI of the brain during the hospital stay were essentially normal. Neuropsychological testing revealed normal cognitive function.

Case 2

Clinical examination revealed as the only finding scars on both knees but was otherwise unremarkable (figure 1). In particular, there were no other signs of a skin disease and no other wounds. Neurological examination was normal.

An ECG showed a normal rhythm with sinus origin and normal depolarisation and repolarisation. Holter monitoring over 48 h was unremarkable. Laboratory findings including haemoglobin, sodium, potassium, calcium and creatinine were normal. TSH and thyroid hormone levels showed a slight hyperthyroid state of function despite the regular intake of carbimazole. An orthostatic test for 12 min showed normal changes in the blood pressure and heart rate. Taking into account the classical presentation as discussed below, the absence of red flags and the long and stable course of the disease, no further testing was performed.

DIFFERENTIAL DIAGNOSIS

In summary, they were two postmenopausal female patients suffering from recurrent falls. As they had never lost consciousness and never experienced any perceptions or residues it would be incorrect to call these patients' troubles syncopes of any cause and it would be inappropriate to suspect seizures. Rather, it might be called 'drop attacks'.

Patients with drop attacks are rare and the differential diagnosis is limited. Drop attacks are imaginable in patients with

narcolepsy, patients with atonic seizures, patients with vertebrobasilar insufficiency, patients with claudicatio intermittens spinalis and patients with untreated thyroid hypofunction. None of these disorders was suspected in these women.

Narcolepsy could be excluded because of the lack of any matching symptoms. Atonic seizures are most common in children.¹ There is a short loss of consciousness and dangerous injuries, especially of the head, are an issue.² Vertebrobasilar insufficiency can lead to drop attacks.¹ However, in this context, one would expect more symptoms and signs of brain-stem affection such as vertigo, double vision, hypo- or paraesthesia of the face or head pain. Moreover, these symptoms are usually provoked by rotation or reclination of the head.¹ Claudicatio intermittens spinalis due to a circulatory disorder of the anterior spinal artery should become evident dependent on the intensity of exercise.

That is why one could call our patients' falls 'cryptogenic drop attacks'.

Interestingly, exactly 40 years previously, an article was published about such a disturbance in the *BMJ*.³

A group in England collected a total of 40 female patients over 12 years reporting about falls without warning, without any sensations and without an underlying disease or condition promoting the falls. In 28 women, follow-up was successful.

Their mean age was 45 years; the youngest patient was 19 years, the oldest 69 years. Two-thirds were between 40 and 59 years. Twenty-six of 28 patients only fell forward and 27 only while walking; all of them suffered injuries to their knees; some also suffered injuries to their wrists, nose, face or chest, but none had severe injuries, especially not to the head. Ten were aware of their falling, 16 at least of crashing to the ground. Two did not recall falling but stood up quickly and resumed walking. Most of them suffered 2–12 falls a year.

As to the prognosis, two patients suffered only one fall in all. Seven of 28 stopped falling within the studied period, whereas 19 continued to fall. Twelve were falling between 1 and 10 years, three between 11 and 20 years and four for more than 20 years.

TREATMENT

There exists no established treatment for this disorder as far as we know. Patients could be instructed to wear kneepads. However, both patients rejected this idea for aesthetic reasons. Furthermore, they should be told that the condition is benign and that no risk of progression is known.

Figure 1 The knees of the second patient with erosions and scars of different age on both sides. (A) and (B) are enlarged sections of the picture.

Case 1: According to the treatment of patients with narcolepsy, we tried a treatment with a tricyclic antidepressant experimentally.⁴ We chose amitriptyline 25 mg at bedtime.

Case 2: This patient did not want pharmacological treatment and chose to wait and see.

OUTCOME AND FOLLOW-UP

Both patients and their family doctors were contacted by phone prior to the writing of this article.

Case 1: Follow-up is 3.5 years. The patient stopped taking amitriptyline after a few months; the reason is not clear, and unfortunately, we cannot reconstruct whether it was helpful or not. However, the falls ceased within a maximum of 1–2 years. Two years after being examined at our clinic, she was diagnosed with mild dementia, but until now she lives at home taking care of herself.

Case 2: Follow-up in this case is 2 years. In this time period, thyroidectomy was performed and carbimazole was discontinued because of poor control. Trazodone was started afterwards because of mild depression and discontinued later. Three months after thyroidectomy, she was sent to the hospital's psychiatrist by the surgeon because of the incidental finding of an early-stage thyroid carcinoma in the struma removed (pT1a). There she was diagnosed with an adjustment disorder with depressed mood but rejected further pharmacological treatment.

Neither of these changes had an influence on her falls. The frequency is still one fall every 1–3 months.

DISCUSSION

When examining a patient after falling, it is helpful to differentiate clearly between five possible etiologic categories: common fall, syncope, collapse, seizure and drop attack.

Syncope is defined as a sudden, short loss of consciousness and tonus with spontaneous recovering in the supine position due to a transient impaired cerebral perfusion. It should be differentiated from a prolonged collapse due to a systemic disturbance such as blood loss, sepsis or cerebral haemorrhage. Seizures are another distinct category with their own features.

Therefore, drop attacks make no part of the differential diagnosis of syncope, collapse or seizures. On the other hand, mechanical reasons for the so-called common falls and their predisposing conditions such as Parkinson's disease, osteoarthritis or impaired vision should be considered.

The very condition of our patients, called 'cryptogenic drop attacks', has been described in a case series 40 years previously as a seemingly distinct entity of drop attacks in women with a very characteristic presentation. In French, it is called 'La Maladie des genoux bleus', which means 'the disease with blue knees'. Obviously, a very concise term.

The diagnosis 'cryptogenic drop attacks' should be a diagnosis of exclusion. However, in selected cases with a very typical presentation, we deem it appropriate to limit evaluation to a careful recording of the patient's history, clinical examination, routine

laboratory values (in particular, blood count, sodium, potassium, calcium, glucose and urinalysis), ECG and follow-up.

Although all the patients in the original paper were women and most of them around the fifth and sixth decade of age, it was not possible to see a direct connection to the menopause.³ Therefore, and because it is not known whether or not hormone substitution would be of any benefit, we did not check the sex hormone levels in our patients.

We have not found additional literature and even case reports are lacking. However, as the above example indicates, the disturbance can be found. Interestingly, in all of the cases of the original series as well as in our two cases, it was impossible to find a causal factor. They are cryptogenic drop attacks—until now. Acknowledgment of this fact—and one look at the patient's knees—may prevent extensive and expensive investigations.

Learning points

- ▶ Not every fall is a syncope.
- ▶ Drop attacks make *no* part of the differential diagnosis of syncope, collapse or seizures due to their own peculiar features.
- ▶ Cryptogenic drop attacks occur only in women, typically in the fifth and sixth decade of life, only while walking, and they lead mainly to contusion of the knees, embarrassment and uncertainty. Prognosis is otherwise good.
- ▶ Alteration of consciousness, prodromes, residues, severe injuries or abnormalities in clinical examination, in routine laboratory findings or in the ECG argue against the diagnosis.
- ▶ In general, the diagnosis 'cryptogenic drop attacks' should be a diagnosis of exclusion.

Contributors RB contributed to taking the patients' histories, clinical examination, diagnosis, follow-up and writing of the article. MS performed a review of the cases and gave approval to the final diagnosis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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