Collapse in a 79-year-old: a rare case of amyloid tumour of the pelvis

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Abstract

A 79-year-old man presented to accident and emergency with collapse, unable to bear weight on his left leg. Computed tomography revealed a large isolated lesion (28 × 12 × 8 cm) extending from the pelvis into the abdomen, affecting the left lumbosacral nerves. Further investigations showed that the mass contained amyloid protein. With no evidence of systemic amyloidosis or malignancy a diagnosis of amyloidoma/amyloid tumour was made. This is the largest amyloid tumour reported in the literature to date. There is limited but conflicting evidence regarding the pathophysiology, management and prognosis of amyloidoma. Clearly amyloidomas are rare, but patients can present acutely and may have a poor prognosis, especially when the tumour is of considerable size.

Keywords: amyloid tumour, amyloidoma, plasmacytoma, elderly

Case report

A 79-year-old man presented to accident and emergency (A&E) with collapse, unable to weight bear on his left leg. He complained of pain on the medial aspect of the left leg, which was worse on extending the hip. There was no recent history of trauma or loss of continence. He had a 25-year history of type 2 diabetes, controlled by insulin, and a 3-year history of late-onset asthma. The patient lived alone with domestic support from a neighbour and had no physical complaints prior to presentation to A&E.

On examination he was alert, afebrile, hypertensive and obese. There was no back tenderness, but there was asymmetry in the flank region (left side larger than right). Sensation to light touch was diminished in the L2–3 dermatome of the left leg. Motor strength of left hip flexion and knee extension was decreased (3/5). The left patellar reflex was absent. All other neurological testing was normal, including the right leg, which was unaffected.

Based on the history and examination a lesion affecting the lumbosacral plexus was suspected at the L3/4 level. Computed tomography (CT) revealed a singular, lobulated homogenous mass infiltrating the left iliac crest (shown in Figure 1). This measured 28×12×8 cm and extended rostrally into the abdomen. Laboratory studies, including full blood count, were within normal limits.

A working differential diagnosis included chondrosarcoma, plasmacytoma/lymphoma or a secondary malignancy. Two separate needle biopsies were inconclusive, yielding amorphous necrotic material. Sections obtained from an open biopsy were positive for amyloid with focal areas of congophilia (with apple-green birefringence). Immunohistochemistry confirmed this and classified the amyloid as a derivative of immunoglobulin light chain, or AL amyloid. Although some cells found in the section were not obviously plasmacytoid in morphology they did react for VS38C (a plasma cell marker), suggesting the possibility of an underlying plasma cell malignancy. Further tests (including echocardiography, skeletal bone scan and urine electrophoresis) showed no evidence of systemic amyloidosis or bone malignancy. A diagnosis of amyloidoma was made.

The initial treatment options were debulking surgery, chemotherapy and/or radiotherapy. Surgery to remove this mass would have necessitated either a massive reconstruction or total amputation (hemi-pelvectomy) as the stability of the pelvis was affected. After consultation with a multidisciplinary team and in accordance with the patient’s wish not to undergo surgery, a course of palliative radiotherapy was offered. Unfortunately the patient’s health declined before this could be started. He was readmitted with sepsis. A follow-up CT showed necrosis within the amyloid tissue and the patient died 3 months after initial diagnosis. A post-mortem examination was not performed.
Discussion

Amyloidoma is a rare manifestation of amyloidosis where abnormal glycoprotein, usually light chain immunoglobulin (AL) or less commonly serum protein A (AA), is deposited locally, forming a solitary mass. Amyloidomas have been reported in a variety of anatomical sites including the gastrointestinal tract [1], central and peripheral nervous system [2], soft tissue [3] and bone. In a review of 34 cases of amyloidoma of the bone the mean age at diagnosis was 57 years (range 27–78), the duration of symptoms ranged from 1 month to 7 years, and the size of the tumour was, on average, 10 cm (range 4–15 cm) [4]. Although Krishnan et al. [5] reported a large retroperitoneal amyloidoma (8 × 12 cm), this case is the largest reported amyloidoma in the literature. Thus far, there are only three previous reports of amyloidoma affecting the pelvis. The first case (58-year-old) presented with pathological fracture [6] while the second tumour (73-year-old) was found incidentally during a follow-up for prostate cancer [7]. The third reported case was in a 40-year-old, presenting as a painless swelling over the left hip, noticed and monitored by the patient for over 11 months [8]. The case discussed here was an acute neurological presentation of a chronic problem that must have been evolving over months, even years, which went unnoticed.

The acute nature of the history and the size of the tumour initially suggested a malignant process of either bone or soft tissue but tissue sampling was necessary for diagnosis. As in this case and others [4, 6–8], histological evidence from needle biopsy was insufficient to make a diagnosis and required open core biopsy and immunohistochemistry studies. Repeated poor quality sections from needle biopsy, yielding amorphous acellular material, may be the first indication of an underlying process involving amyloid.

Management of this condition was a challenge due to the paucity of documented treatment options. For small masses, especially those affecting the vertebrae, surgical debulking has been successful in removing the space-occupying effects of the amyloidoma [10]. Surgery would have been too disfiguring. The alternative treatment option was radiotherapy to decrease the bulk of the tumour and possibly interrupt a potential underlying malignancy of the bone. Chemotherapy has been used in cases where a definite diagnosis of an underlying myeloma has been shown [5], which was not the case here.

The pathophysiology of amyloidoma is unclear. Some reports suggest that the amyloid is produced within an immunocytic neoplasm such as a plasmacytoma [3, 4]. This underlying malignancy may not be found at diagnosis due to ‘burn out’, where the neoplasm has been overwhelmed by amyloid deposits [9]. In a review of 14 cases of soft tissue amyloidoma, 80% of patients with type AL amyloidomas were later found to have a peripheral B-cell neoplasm (plasmacytoma/lymphoplasmacytoid lymphoma) with an overall poor prognosis [5]. This is in agreement with Pambuccian et al. [4] where the prognosis of patients with AL amyloidoma is given as 8 months to 3 years with most dying of local or general complications. However, there have been cases showing excellent prognosis with no evidence for increased mortality [10]. This discrepancy may be associated with the type of amyloid deposited, with type AL amyloidomas having a poorer prognosis than type AA [5]. In addition, prognosis is likely to be affected by the size and location of the tumour. Surgical management of small tumours without underlying malignancy has been shown to be curable [10], while management of large tumours tends to be supportive, as in this case.

Interpretation of factors influencing prognosis in amyloidomas is difficult due to the limited evidence base, lack of adequate follow-up and the retrospective approach in the reported literature. This case highlights the difficulty in diagnosis, management and prognosis of a rare tumour that can present in later life.

Key points

- Amyloidoma is an isolated collection of glycoprotein deposited extracellularly in the tissues, forming a solitary mass (amyloid tumour).
- Diagnosis of amyloidoma requires an adequate tissue biopsy sample with the use of congo-red stain and immunohistochemistry studies.
- Management of large tumours is usually supportive.
- Amyloidoma of the bone is thought to be associated with an underlying plasmacytoma.

References

Buried Bumper Syndrome complicated by intra-abdominal sepsis

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Abstract

There is growing evidence that enteral feeding tubes are associated with increased mortality and complication rates in patients with advanced dementia. Buried Bumper Syndrome is an uncommon, but well documented complication of PEG placement. Our case report reinforces this recognised risk of PEG feeding in an elderly, cognitively impaired patient.

Keywords: percutaneous endoscopic gastrostomy, dementia, buried bumper syndrome

Introduction

Percutaneous Endoscopic Gastrostomy (PEG) is used increasingly for long-term enteral support in patients with dementia. However, numerous complications have been reported since its introduction in 1980 [1]. Buried Bumper Syndrome (BBS) is an uncommon, but well documented complication of PEG placement. We report a case of BBS complicated by a large intra-abdominal abscess.

Case report

An 81-year-old lady with advanced dementia was admitted to our hospital from a nursing home with vomiting and rigors. A PEG had been in situ for 3 years. Examination revealed a large, tender, right-sided abdominal mass. She was tachycardic and pyrexial with raised inflammatory markers. Ultrasound abdomen revealed a large intra-peritoneal abscess. Cultures grew enterococcal species and she was commenced on intravenous antibiotics. Gastroscopy and CT abdomen revealed migration of the PEG internal bumper out of the stomach. The patient was referred to the surgeons for urgent exploration and removal of PEG. She died two days after the procedure following a brainstem infarction.

Discussion

BBS is a serious complication of PEG tube insertion first described in 1988 [2, 3] and reported to occur in 0.3–2.4% of patients [4]. It is a late complication occurring up to 3 years post PEG insertion [5], but has been described at 21